Cone versus conventional repair for Ebstein's anomaly

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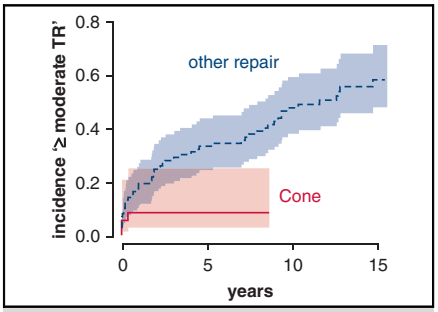
ABSTRACT

Objectives: We aimed to investigate tricuspid valve function and adverse events after conventional repair and valve replacement for Ebstein's anomaly and compare them with cone repair.

Methods: The medical records of 151 patients (mean age, 25 years; 62% were female) who underwent operation in a single center from 1985 to 2018 were retrospectively analyzed. To determine tricuspid valve regurgitation during follow-up, serial echocardiographic examination was used (n = 2397, tricuspid regurgitation grades were graphed for every patient).

Results: Thirty-nine patients underwent cone repair, 107 patients underwent other repair techniques, and 5 patients underwent valve replacement. The operative mortality was 1.3% (n = 2). Failed valve repair (defined as in-hospital death, conversion to replacement, or in-hospital reoperation) was less frequent after cone repair than after other repair techniques (5%, n = 2 vs 20%, n = 21, $P = .039$). Mean follow-up was 12.3 years (cone repair: 3.7 years). The 5-year cumulative incidence of moderate or greater recurrent tricuspid regurgitation was lower after cone repair than after other repair techniques (8% vs 32%, $P = .03$). Among the patients undergoing other repair techniques, the 15-year cumulative incidence of moderate or greater recurrent tricuspid regurgitation, severe tricuspid regurgitation, and reoperation was 58%, 37%, and 31%, respectively. During follow-up, 18 patients died (13 of cardiac and 5 of noncardiac causes). Among patients who died of cardiac causes, 10 of 13 had all 3 characteristics—moderate or greater tricuspid regurgitation, atrial fibrillation, and New York Heart Association classification III and IV—at their last medical evaluation.

Conclusions: Before cone repair, recurrent tricuspid regurgitation was considerable. Cone repair provided a higher rate of successful repair and a lower incidence of moderate or greater recurrent tricuspid regurgitation at the midterm follow-up. (J Thorac Cardiovasc Surg 2020;160:1545-53)



TR after cone repair (red) and other repair techniques (blue).

CENTRAL MESSAGE

Cone repair provided a higher rate of successful repair and a lower incidence of moderate or greater recurrent insufficiency compared with former repair procedures.

PERSPECTIVE

In the past, residual TR and redo surgery were frequent after TV repair for Ebstein's anomaly. The recently introduced cone repair provides a more reliable valve repair.

See Commentaries on pages 1554, 1556, and 1557.

Ebstein's anomaly is a rare malformation of the tricuspid valve (TV) that accounts for 1% of all congenital heart defects.[1] Its main feature is the failure of delamination of the TV from the underlying myocardium. Consequently, the leaflets are adherent to the myocardium, especially at the septum and posterior wall. The TV is often regurgitant to varying degrees, and the annulus is grossly dilated.

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Abbreviations and Acronyms
CI = confidence interval
NYHA = New York Heart Association
RV = right ventricle
TR = tricuspid regurgitation
TV = tricuspid valve

Original TV reconstruction techniques for Ebstein's anomaly were based on ventricular plication, with the aim to bridge leaflet gaps and to decrease the massively dilated tricuspid annulus.[2] Derived from an innovative operation technique from Carpentier and colleagues,[3] da Silva and colleagues[4] developed the cone technique in 1993. In the cone repair, all 3 leaflets are almost completely detached from the ventricular wall except at their hinge point, that is, displaced toward the outflow tract. The right ventricle (RV) is then longitudinally plicated (folded) at the posterior wall to exclude the atrialized portion of the RV, thus reducing the volume of the RV and the diameter of the annulus. The leaflets are rotated and reattached to the morphologic tricuspid annulus. All 3 commissures are sutured together longitudinally. The resulting reconstructed valve resembles a cone that covers 360° of the annulus and is directed toward the ventricle. Within the past decade, centers performing a large number of surgical procedures for Ebstein's anomaly have almost simultaneously adopted this technique.[5-10]

Valve competence after surgical repair for Ebstein's anomaly has often been unsatisfactory, resulting in a high incidence of reoperation in the long term (42%),[11] leading eventually to right heart failure, atrial fibrillation, and finally death. In this study, we aimed to investigate TV function and adverse events after conventional repair of Ebstein's anomaly and compare the results with cone repair.

MATERIALS AND METHODS

This study was approved by the Ethics Committee of the Technical University Munich (project number 554/18). The Ethics Committee waived the need for informed consent from the parents or patients because of the lack of any patient identifiers and the retrospective nature of the study. We retrospectively analyzed the data of patients with Ebstein's anomaly who underwent TV surgery at our center from January 1985 to October 2018. The cone repair was introduced in 2010.[6] Patients who underwent surgery before 1985 were not included because echocardiographic examinations were not performed at that time. Exclusion criteria were as follows: (1) prior TV surgery at another institution, (2) surgery as a neonate, and (3) congenitally corrected transposition of the great arteries.

We reviewed the medical records from hospital stays and from visits at the outpatient clinic and the reports from referring cardiologists. Preoperative characteristics, procedural data, and postoperative data on tricuspid regurgitation (TR) and adverse events were collected.

All available reports of echocardiographic examinations were reviewed. TR was graded as follows: none, trivial, mild, moderate, or severe according to the examiner's description. The values were displayed graphically against postoperative time for each patient. The cumulative incidence rates of moderate or greater TR and severe TR were determined on the basis of the postoperative time of their first occurrence. If a once-only assessment of moderate/severe TR was followed by multiple assessments yielding TRs

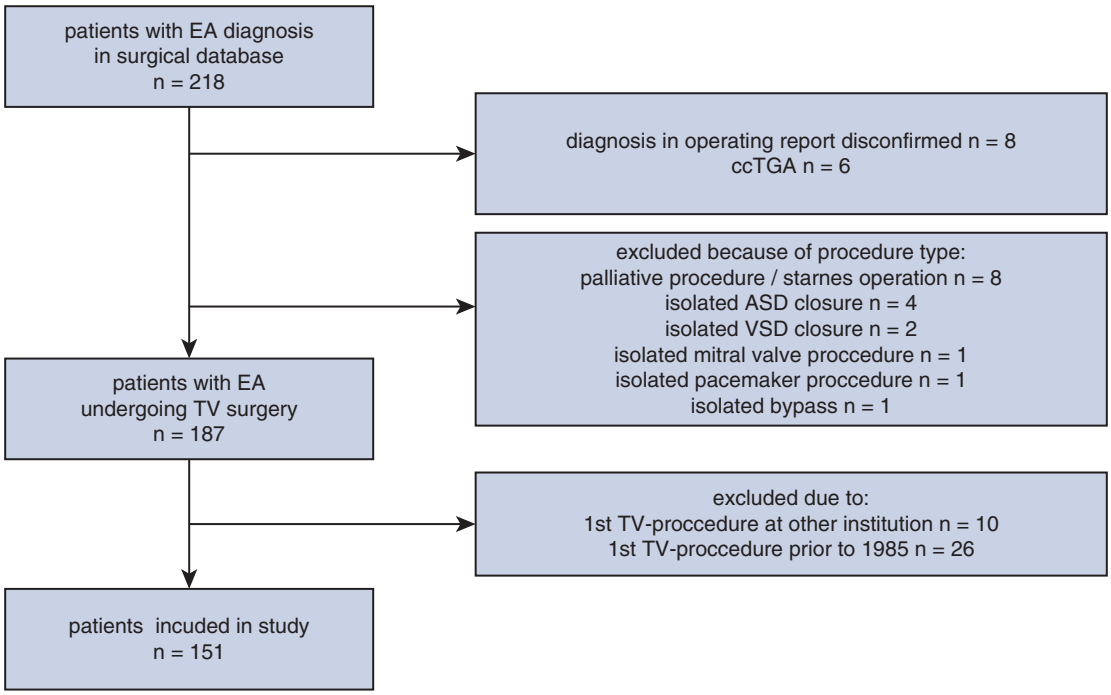


FIGURE 1. Flow diagram of patient inclusion/exclusion. *EA,* Ebstein's anomaly; *ccTGA,* congenitally corrected transposition of the great arteries; *TV,* tricuspid valve; *ASD,* atrial septal defect; *VSD,* ventricular septal defect.

less than moderate, we considered the moderate/severe TR assessment to be an outlier and ignored this event.

For analysis, patients were categorized into 3 groups for analysis, according to the operative technique used at their first TV surgery, as follows: (1) cone repair technique, (2) other repair techniques, and (3) valve replacement. Categoric variables were described as counts and percentages, and continuous variables were expressed as means ± standard deviations or medians and ranges. The 95% confidence intervals (CIs) were reported. The Kaplan–Meier method was used for survival analysis. Other time-to-event analysis was performed using the cumulative incidence function. Comparisons between groups were performed using the Student *t* test for normally distributed variables, the Wilcox rank-sum test for not normally distributed variables, and the Fisher exact test for binominal variables. All statistical calculations were performed using the R statistical computing environment (version 3.5.0).

RESULTS

Among the 218 patients with the diagnosis of Ebstein's anomaly who underwent a surgical procedure at our institution, 151 fulfilled the inclusion criteria for this study (Figure 1). Mean age was 28 years, and 56 patients (37%) were aged less than 18 years. Reoperations and mortality during hospital stay are depicted in Figure 2. In 5 of 151 patients (3%), valve repair was not attempted and the valve was replaced directly. In the remaining 146 patients (97%), a valve repair was attempted (cone repair n = 39, other repair techniques n = 107). The cone repair was performed according to the original description by da Silva and colleagues (Video 1).[5] There were no annuloplasty rings used. A limited interatrial connection was left in all patients who underwent the cone procedure. Descriptions and frequencies of "other repair techniques" are displayed in Online Data Supplements 1 to 4. The majority of these procedures aimed to approximate the anterior leaflet to the septum (Sebening monocusp technique).[12] In all patients undergoing valve replacement, a biological prosthesis was implanted. Of 39 patients undergoing the cone repair, 1 was converted intraoperatively to valve replacement and 1 patient died during the hospital stay. Of 107 patients undergoing other repair techniques, 5 were converted intraoperatively to valve replacement, 15 required a reoperation before hospital discharge (7 re-repair, 8 valve replacement, median time to reoperation: 7 days), and 1 died during the hospital stay. None of the patients who underwent valve replacement died during the hospital stay.

The cone repair technique provided a significantly higher rate of successful repair (defined as no conversion to replacement, reoperation before discharge, or in-hospital mortality) than the other repair procedures (95% vs 80%, respectively; $P = .039$). Median hospital stay was 15 days. At hospital discharge, the final postoperative results were cone repair in 37 patients, another repair in 93 patients, valve replacement in 19 patients, and death in 2 patients. The preoperative characteristics and procedural data of the 3 groups are shown in Table 1. Two patients died during the hospital stay (1.3%) after surgery as

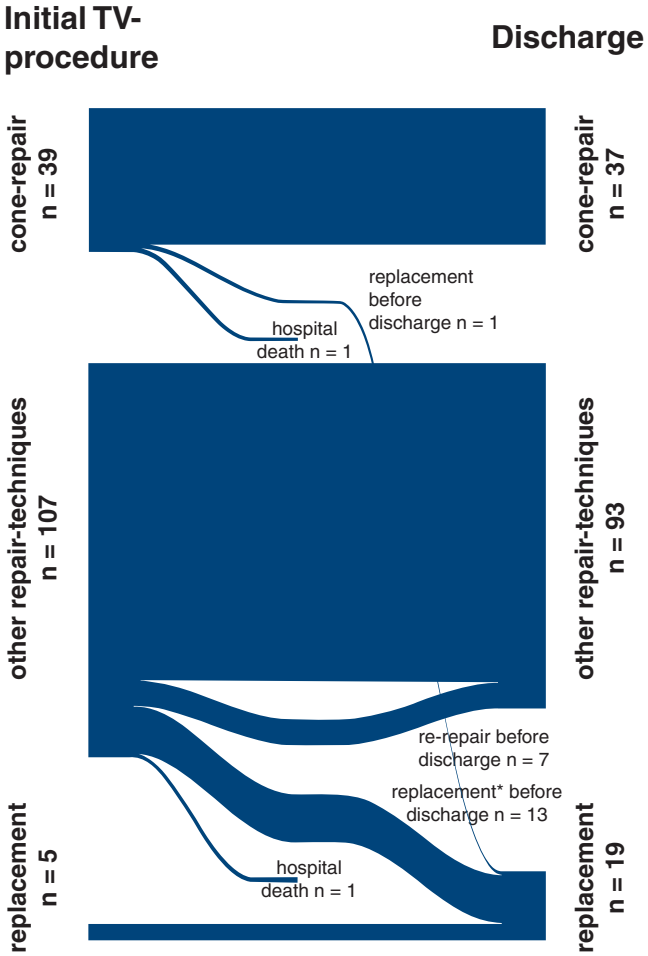
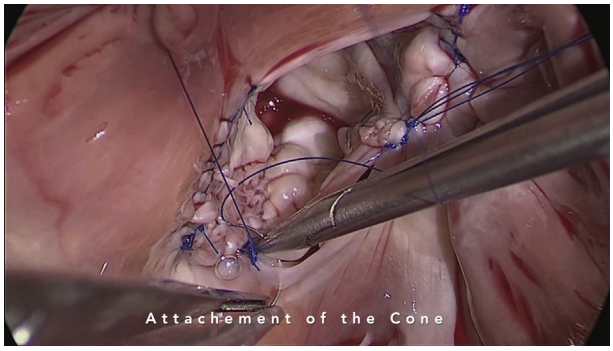


FIGURE 2. River-plot summarizing hospital mortality, reoperation, and conversion from repair to valve replacement before hospital discharge. *Conversion to valve replacement during initial operation or valve replacement in a second operation before discharge. *TV,* Tricuspid valve.

follows: (1) A 49-year-old patient died suddenly 12 days after the repair procedure from hematopericardial tamponade, and (2) a 61-year-old patient died of multiple organ failure 2 weeks after a cone repair. Postoperative valve



VIDEO 1. Cone repair performed in a 53-year-old woman who presented with severe TR. Video available at: https://www.jtcvs.org/article/S0022-5223(20)31251-4/fulltext.

TABLE 1. Preoperative characteristics, procedural data, and in-hospital complications of tricuspid valve surgery for Ebstein's anomaly

	All	Cone repair	Other repair	Replacement
Preoperative characteristics				
Patients (n)	151	38	94	19
Year of operation*,†,‡				
1985-2000	52 (34%)	0 (0%)	52 (55%)	0 (0%)
2000-2010	57 (38%)	0 (0%)	40 (43%)	17 (89%)
2010-2018	42 (28%)	38 (100%)	2 (2%)	2 (11%)
Age (y)	28 ± 19	31 ± 19	26 ± 18	36 ± 21
Gender (female)	90 (60%)	23 (61%)	58 (62%)	9 (47%)
Previous intervention ASD closure	5 (3%)	3 (8%)	2 (2%)	0 (0%)
Previous surgical ASD closure	6 (4%)	3 (8%)	2 (2%)	1 (5%)
Previous stroke	15 (10%)	6 (16%)	9 (10%)	0 (0%)
Wolff–Parkinson–White syndrome	24 (16%)	7 (18%)	15 (16%)	2 (11%)
Previous ablation	28 (19%)	10 (26%)	15 (16%)	3 (16%)
Previous pacemaker implantation	3 (2%)	0 (0%)	2 (2%)	1 (5%)
Anticoagulation before surgery	15 (10%)	5 (13%)	6 (6%)	4 (21%)
Preoperative TR grade				
Moderate	35 (23%)	6 (16%)	26 (28%)	3 (16%)
Severe	110 (73%)	32 (84%)	62 (66%)	16 (84%)
Unknown	6 (4%)	0 (0%)	6 (6%)	0 (0%)
Procedural data				
Cavopulmonary anastomosis	1 (1%)	1 (3%)	0 (0%)	0 (0%)
Cardiopulmonary bypass time*,‡	117 ± 47	149 ± 28	94 ± 43	131 ± 49
Mean aortic clamp time*,†,‡	74 ± 41	117 ± 20	49 ± 30	69 ± 34
Postoperative complications, postoperative management				
Mortality (in hospital)	2 (1.3%)	1 (3%)	1 (1%)	0 (0%)
Use of ECMO*	4 (3%)	4 (11%)	0 (0%)	0 (0%)
Stroke (in hospital)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Pacemaker implantation (in hospital)	14 (9%)	2 (5%)	8 (9%)	4 (21%)

Statistically significant difference between groups are as follows: noted in superscript mark-up: *Significant difference between the cone technique and other repair. †Significant difference between cone technique and replacement. ‡Significant difference between other repair techniques and replacement). All *P* values are specified in the Online Data Supplements 1 to 4. *ASD,* Atrial septal defect; *TR,* tricuspid regurgitation; *ECMO,* extracorporeal membrane oxygenation.

performance was excellent in both patients; TR was less than mild.

Follow-up After Hospital Discharge

Two patients were lost to follow-up immediately after discharge. Follow-up data were available from all the remaining patients (98%) as follows: Life status/follow-up data collected in 2017 and 2018 were available for 129 patients (85%). In 10 patients (7%), follow-up was available but only from before 2010; 7 of these patients were living abroad. A total of 2397 postoperative echocardiographic examinations were available for serial analysis (mean: 16 echocardiographic examinations per patient; 83% were performed at our center, and 17% by a referring cardiologist). For each patient, a figure displaying the serial assessment of TR was created (Online Data Supplements 1-4). Adverse events were obtained from 1935 medical reports.

Figure 3 summarizes the outcome after the cone repair compared with other repair techniques. The incidence of recurrent TV regurgitation and reoperation after TV replacement is shown in Online Data Supplements 1 to 4. The incidence rate of recurrent TR moderate or greater was lower after cone repair than after the other repair techniques ($P = .031$). A total of 34 patients developed recurrent severe TR during the follow-up. Thirty-one of these patients exhibited moderate TR in their previous echocardiographic examination. The median time from the first occurrence of moderate TR to the first occurrence of severe TR was 4.7 years. The incidence rates of severe TR and reoperations after cone repair did not differ significantly ($P = .29$ and $P = .14$). The reoperations performed after valve repair consisted of re-repair (n = 15; 56%) and valve replacement (n = 12; 44%). The reoperations performed after valve replacement consisted of re-replacement (n = 2; 50%) and interventional valve-in-valve implantation (n = 2; 50%). Two patients underwent heart transplantation during follow-up. Three of 14 pacemaker implantations performed during follow-up were associated with valve replacement. Table 2 displays all documented postoperative events.

Figure 4 shows the Kaplan–Meier estimation for survival after hospital discharge. Survival did not differ significantly among patient groups. Table 3 displays the detail about the

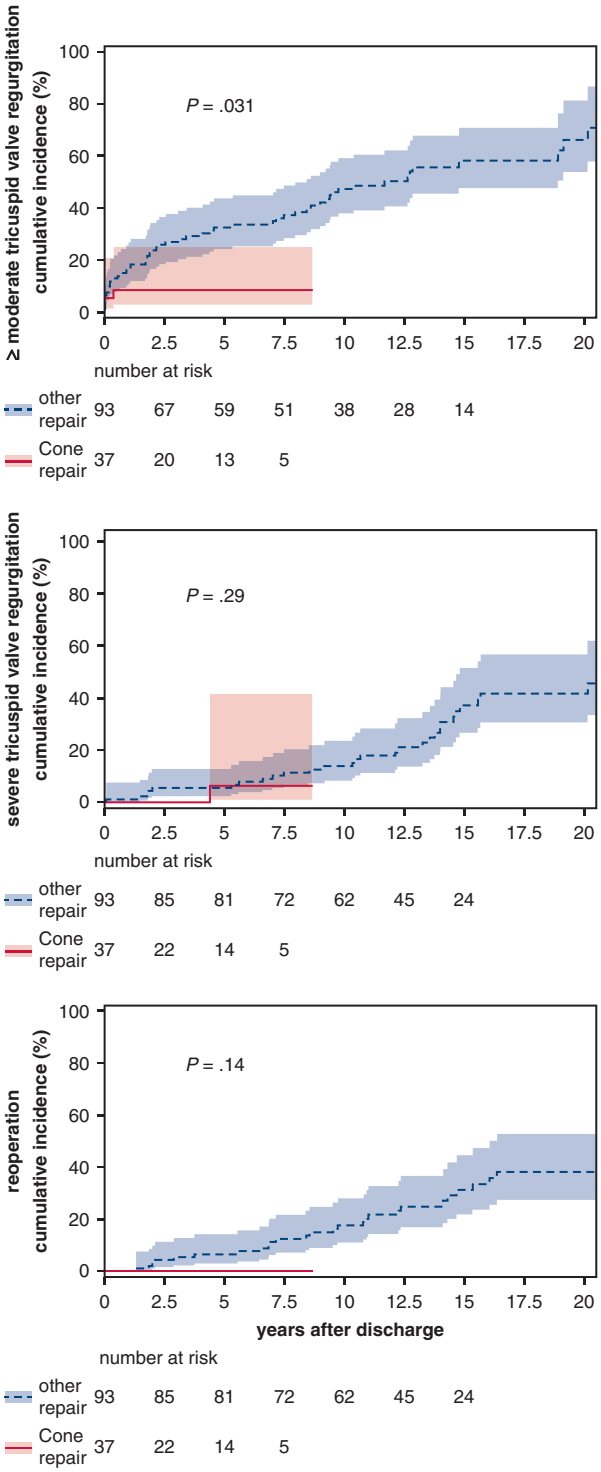


FIGURE 3. Cumulative incidence of moderate or greater TR, severe TR, and reoperation after cone repair compared with other repair techniques. The 95% CIs are plotted as shades.

18 patients who died after hospital discharge. Thirteen patients died of cardiac causes, and 5 patients died of noncardiac causes. The mean age at death was 49 ± 18 years. Ten of 13 deaths from cardiac causes were associated with reoperation or with all 3 of the following characteristics: TR moderate or greater, atrial fibrillation, and New York Heart Association (NYHA) III to IV. Sudden death despite sinus rhythm, good TV function, and NYHA I occurred in 1 patient. Of note, 2 of 4 deaths among patients who underwent TV replacement were noncardiac and probably not associated with the TV procedure. This puts the lower survival of the group of patients who underwent TV replacement (Figure 4) into perspective.

DISCUSSION

Conventional repair techniques for Ebstein's anomaly before the cone repair reduced TR and resulted in an excellent survival in hospital. However, 20% of the patients required a conversion to replacement during the initial procedure or underwent repeat surgery before hospital discharge. In addition, a significant number of these patients developed TR during follow-up. The recently introduced cone repair was followed by consistently good tricuspid function over an 8-year follow-up period with significantly fewer patients with moderate or greater tricuspid insufficiency than the other surgical methods. In addition, the cone repair was followed by a higher rate of operative success than the other procedures, as reflected by fewer conversions to valve replacement or reoperations before hospital discharge. Therefore, we think that the cone repair should be the primary treatment for patients with Ebstein's anomaly.

To our knowledge, this is the first study using serial echocardiographic data to assess TV function after various surgical procedures for Ebstein's anomaly and compare those with valve function after the cone repair. Previous reports assessed postoperative valve function by echocardiographic examination at discharge or last follow-up[9,13,14] or by freedom from reoperation.[15] However, to detect changes in valve function after surgical procedures for Ebstein's anomaly, sequential assessments of valve function is paramount.[16] Furthermore, the incorporation of multiple measurements by different cardiologists reduces subjectivity and enables correction of outlier values. The analysis of only freedom from reoperation overestimates the quality of valve function, because it does not address the following situations: delay between first signs of valve dysfunction and reoperation, patients who cannot undergo reoperation despite valve dysfunction, and patients with moderate TR but without an indication for reoperation.

The serial echocardiographic analysis showed that repair techniques other than the cone repair were followed by a high incidence of TR moderate or greater within the first few years after operation, that is, 32% after 5 years and 47% after 10 years. The incidence of severe TV regurgitation at 5 years was 8%, but increased to 19% at 10 years and 47% at 20 years. In contrast, 5 years after

TABLE 2. Adverse events during follow-up after tricuspid valve surgery in patients with Ebstein's anomaly

		All	Cone repair	Other repair	Replacement
Patients (hospital survivors)		149	37	93	19
Mean follow-up*,†,‡		12.3 ± 8.7	3.7 ± 2.7	16.3 ± 8	9.6 ± 5.6
Patient-y		1832	138	1512	182
Echo per patient (mean)*,†		16	9	19	17
Deaths during follow-up	n (%)	18 (12%)	0 (0%)	14 (15%)	4 (21%)
Survival (Kaplan–Meier estimator)	at 5 y	97.7	100.0	97.8	94.1
	at 10 y	93.7	NA	94.1	87.8
	at 20 y	81.5	NA	83.5	NA
Reoperations	n (%)	32 (21.%)	0 (0%)	28 (30%)	4 (21%)
Cumulative incidence	at 5 y	0.05	0.00	0.07	0.06
	at 10 y	0.16	NA	0.18	0.19
	at 20 y	0.36	NA	0.38	NA
TV regurgitation moderate or less*		64 (43%)	3 (8.1%)	54 (58%)	7 (37%)
Cumulative incidence	at 5 y	0.30	0.08	0.32	0.31
	at 10 y	0.44	NA	0.47	0.37
	at 20 y	0.67	NA	0.66	NA
Severe TV regurgitation		34 (23%)	1 (2.7%)	29 (31%)	4 (21%)
Cumulative incidence	at 5 y	0.08	0.06	0.08	0.13
	at 10 y	0.18	NA	0.14	0.19
	at 20 y	0.45	NA	0.42	NA
Catheter ablation		27 (18%)	2 (5.41%)	22 (24%)	3 (16%)
Cumulative incidence	at 5 y	0.11	0.08	0.11	0.18
	at 10 y	0.17	NA	0.19	0.18
	at 20 y	0.27	NA	0.28	NA
Prescription of anticoagulation	35 (23%)	3 (8.1%)	25 (27%)	7 (37%)	
Cumulative incidence	at 5 y	0.11	0.09	0.09	0.27
	at 20 y	0.34	NA	0.31	NA
Pacemaker implantation during follow-up	14 (9.4%)	1 (2.7%)	10 (11%)	3 (16%)	
Cumulative incidence	at 5 y	0.06	0.03	0.04	0.16
	at 20 y	0.16	NA	0.15	NA
Other events					
Endocarditis n (ppy %)		1 (0.06%)	0 (0%)	1 (0.07%)	0 (0%)
Ischemic stroke n (ppy %)		4 (0.22%)	1 (0.80%)	3 (0.20%)	0 (0%)
Intracranial bleeding n (ppy %)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	
Other major bleeding n (ppy %)‡	1 (0.05%)	0 (0%)	0 (0%)	1 (0.55%)	

Statistical significant difference between groups is as follows: *Significant difference between cone technique and other repair. †Significant difference between cone technique and replacement. ‡Significant difference between other repair techniques and replacement. All *P* values are specified in Online Data Supplements 1 to 4. *NA,* Not available; *TV,* tricuspid valve; *ppy,* per patient year.

the cone repair, moderate or greater TV insufficiency occurred in only 8% and severe TV insufficiency in only 6%. The rate of severe recurrent TR during the first 5 years of follow-up was relatively low for all repair techniques. This is probably attributable to the fact that if a patient exhibited moderate to severe TR postoperatively, a re-repair or replacement was performed before hospital discharge. In contrast, the presence of mild (to moderate) TR postoperatively was often accepted, because a re-repair did not ensure an improvement of the result. This was a common situation before the cone era. The presence of moderate TR is relevant for several reasons: First, we identified moderate TR as a risk factor for the progression to severe TR. Second, we found that most patients who die of a cardiac cause had at least moderate TR, in combination with atrial fibrillation and NYHA classification III and IV. Third, moderate TR restricts physical activities, according to the recommendations of current guidelines.[17] Fourth, residual TR may promote later atrial arrhythmias in the long-term.

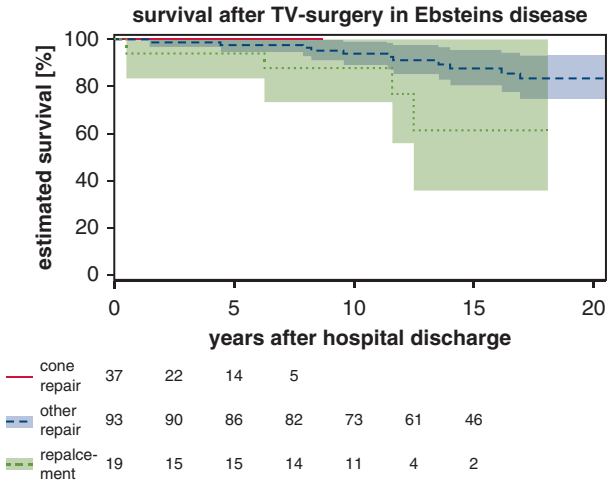


FIGURE 4. Kaplan–Meier survival curves of patients who underwent TV surgery for Ebstein's anomaly (operative mortality not included). The 95% CIs are plotted as shades. *TV,* Tricuspid valve.

Attenhofer and colleagues[18] recently reported the important role of right heart failure in premature death in Ebstein's anomaly after TV surgery. In a cohort composed of 968 patients, they found that the cumulative incidence of sudden death was 10.8% 20 years after TV surgery. Patients with heart failure had a 6-fold increased risk of sudden death.[18] In the current study, sudden death of patients in otherwise good condition was extremely rare; only 1 patient died suddenly despite good valve function, sinus rhythm, and stable clinical condition. The pathophysiologic sequence behind these deaths could be recurrent TR moderate or greater after repair, progressive enlargement of the RV and atrium,[19] development of atrial fibrillation, and symptoms of heart failure. The newly introduced cone repair was followed by a relatively low rate of recurrent TR during a follow-up of 8 years. We speculate that the lower incidence of TR after cone repair reduced the progression to heart failure.

TABLE 3. Cause of death after tricuspid valve surgery in patients with Ebstein's disease

Operation	Time operation, death (y)	Age at operation (y)	Redo	Cause of death	RV function	TR grade	ECG	NYHA
Monocusp	12 d	49		Hematopericardium				
Cone	14 d	61		RV failure, ECMO		-	-	-
Replacement	0.5	66		Sudden death	Reduced	Mild	Atrial fibrillation	IV
Monocusp	1.5	23	Y	VT in rehabilitation after redo	Unknown	None	Sinus rhythm	I
Monocusp	4.5	13	Y	Sudden death	Reduced	Moderate	Atrial fibrillation	n
Monocusp	8	1		Sudden death, fever	Reduced	Severe	Atrial fibrillation	IV
Monocusp	8	38		Heart failure	Reduced	Severe	Atrial fibrillation	IV
Monocusp	10	31		Sudden death	Reduced	Moderate	Atrial fibrillation	IV
Bicuspid	11	52		Sudden death	Mildly reduced	Moderate	Intermediate atrial fibrillation	III
Replacement	12	47		Sudden death	Normal	None	Intermediate atrial fibrillation	III
Monocusp	12	57		Heart failure	Unknown	Moderate	Atrial fibrillation	IV
Monocusp	14	31		Sudden death	Unknown	None	Sinus rhythm	I
Monocusp	17	51		Heart failure	Unknown	Severe	Atrial fibrillation	IV
Monocusp	23	24	Y	Death in hospital after	Unknown	-	-	-
Monocusp	29	32	Y	In hospital after reoperation, RV failure	Reduced	None	-	-
Replacement	6	40		Early dementia	Normal	Severe	Sinus rhythm	I
Replacement	12	53		Abdominal aortic aneurysm	Reduced	Mild	Atrial fibrillation	I
Bicuspid	14	63		Pancreas-carcinoma	normal	Moderate	Atrial fibrillation	II
Monocusp	1 (5)	18		Accident (on Htx list)	Reduced	Severe	Atrial fibrillation	IV
Monocusp	22	17	Y	Colon-carcinoma	Mildly reduced	Moderate	Sinus rhythm	II-III

Redo means patient underwent reoperation on the TV during follow-up. *RV,* Right ventricle; *TR,* tricuspid regurgitation; *ECG,* electrocardiogram; *NYHA,* New York Heart Association; *ECMO,* extracorporeal membrane oxygenation; *VT,* ventricular tachycardia; *Htx,* heart transplantation.

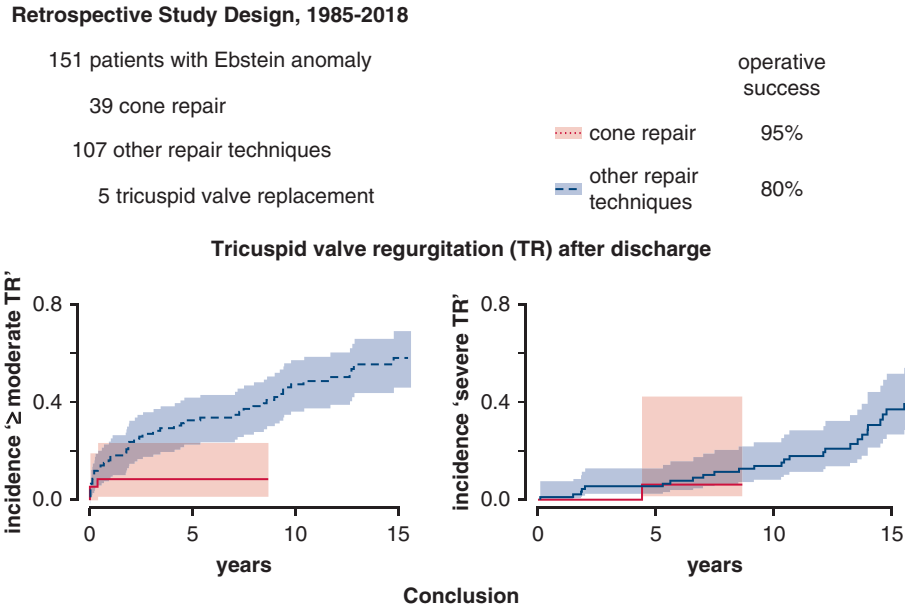


FIGURE 5. Recurrent TR after cone repair (red) and other repair techniques (blue) was compared using serial echocardiographic examinations.

Because most patients in the present study underwent valve repair, only limited conclusions about TV replacement can be drawn from our findings. Because of degeneration of the bioprosthesis, a progressive increase of tricuspid insufficiency was seen after valve replacement that began early after the initial replacement. In addition, the pacemaker implantation rate and the incidence of atrial arrhythmia were higher and long-term survival was lower than after conventional and cone repair techniques. Likewise, a recent study from our institution of 51 patients with congenital heart defects who underwent TV replacement showed a freedom from reoperation rate at 5 and 10 postoperative years of only 86% and 81%, respectively, and from prosthesis dysfunction of only 66% and 58%, respectively. Valve implantation at an age younger than 16 years was associated with even earlier dysfunction.[20] These findings suggest that all attempts should be made to repair rather than replace the TV, especially in younger patients. However, in older patients with an enlarged RV and impaired RV function, the risk of postoperative RV failure is substantial.[8] In such patients, a bioprosthesis may be preferred over complex valve repair such as the cone operation. For those patients, endovascular implantation has become important in patients with prosthesis dysfunction.

Study Limitations

This study was a retrospective single-center study and could not report comprehensively on all the repair techniques used elsewhere for Ebstein's anomaly. The cone repair was performed more recently than the other repair techniques. Comparing techniques from different eras always has limitations, because many factors change in the course of time. Furthermore, the follow-up length of the cone group was consequently significantly shorter compared with the other repair techniques, and there were only 14 patients with a cone repair who had a follow-up longer than 5 years. Only written findings of the echocardiographic examinations were reviewed, not the actual images. These findings are prone to interobserver variation. Objective measurements such as the size of the vena contracta were rarely documented and therefore not available for analysis. Although we attempted to obtain echocardiographic data as complete as possible, there are gaps within the serial echocardiographic evaluations, and the detection of TR sometimes may have been delayed. Thus, because of delayed detection, the time-to-event analysis might have underestimated the "true" incidence of TR.

CONCLUSIONS

Recurrent and progressive TR is frequently seen after conventional repair of Ebstein's anomaly. In addition, 20% of the patients required conversion to TV replacement or other repeat surgery before hospital discharge. The recently introduced cone repair yields consistent tricuspid function over an 8-year follow-up period (Figure 5). Although long-term results after the cone procedure are still pending, we think that the cone repair should be the primary treatment for patients with Ebstein's anomaly.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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References

1. Correa-Villaseñor A, Ferencz C, Neill CA, Wilson PD, Boughman JA. Ebstein's malformation of the tricuspid valve: genetic and environmental factors. *Teratology*. 1994;50:137-47.
2. Hardy KL, May IA, Webster CA, Kimball KG. Ebstein's anomaly: a functional concept and successful definitive repair. *J Thorac Cardiovasc Surg*. 1964;48:927-40.
3. 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.
4. da Silva JP, Baumgratz JF, da Fonseca L, Franchi SM, Lopes LM, Tavares GMP, et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results. *J Thorac Cardiovasc Surg*. 2007;133:215-23.
5. Dearani JA, Said SM, O'Leary PW, Burkhart HM, Barnes RD, Cetta F. Anatomic repair of Ebstein's malformation: lessons learned with cone reconstruction. *Ann Thorac Surg*. 2013;95:220-8.
6. Lange R, Burri M, Eschenbach LK, Badiu CC, da Silva JP, Nagdyman N, et al. da Silva's cone repair for Ebstein's anomaly: effect on right ventricular size and function. *Eur J Cardiothorac Surg*. 2014;48:316-21.
7. Liu J, Qiu L, Zhu Z, Chen H, Hong H. Cone reconstruction of the tricuspid valve in Ebstein anomaly with or without one and a half ventricle repair. *J Thorac Cardiovasc Surg*. 2011;141:1178-83.
8. Mrad Agua K, Burri M, Cleuziou J, Beran E, Meierhofer C, Nagdyman N, et al. Preoperative predictability of right ventricular failure following surgery for Ebstein's anomaly. *Eur J Cardiothorac Surg*. 2018;55:1187-93.
9. Vogel M, Marx GR, Tworetzky W, Cecchin F, Graham D, Mayer JE, et al. Ebstein's malformation of the tricuspid valve: short-term outcomes of the "cone repair" versus conventional surgery. *Congenit Heart Dis*. 2012;7:50-8.
10. Ibrahim M, Tsang VT, Caruana M, Hughes ML, Jenkyns S, Perdreau E, et al. Cone reconstruction for Ebstein's anomaly: patient outcomes, biventricular function, and cardiopulmonary exercise capacity. *J Thorac Cardiovasc Surg*. 2015;149:1144-50.
11. Badiu CC, Schreiber C, Hörer J, Ruzicka DJ, Wottke M, Cleuziou J, et al. Early timing of surgical intervention in patients with Ebstein's anomaly predicts superior long-term outcome. *Eur J Cardiothorac Surg*. 2010;37:186-92.
12. Schmidt-Habelmann P, Meisner H, Struck E, Sebening F. Results of valvuloplasty for Ebstein's anomaly. *Thorac Cardiovasc Surg*. 1981;29:155-7.
13. Hetzer R, Hacke P, Javier M, Miera O, Schmitt K, Weng Y, et al. The long-term impact of various techniques for tricuspid repair in Ebstein's anomaly. *J Thorac Cardiovasc Surg*. 2015;150:1212-9.
14. Holst KA, Dearani JA, Said S, Pike RB, Connolly HM, Cannon BC, et al. Improving results of surgery for Ebstein anomaly: where are we after 235 cone repairs? *Ann Thorac Surg*. 2018;105:160-8.
15. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA, et al. Functional status after operation for Ebstein anomaly. *J Am Coll Cardiol*. 2008;52:460-6.
16. Capodanno D, Petronio AS, Prendergast B, Eltchaninoff H, Vahanian A, Modine T, et al. Standardized definitions of structural deterioration and valve failure in assessing long-term durability of transcatheter and surgical aortic bioprosthetic valves: a consensus statement from the European Association of Percutaneous Cardiovascular Interventions (EAPCI) endorsed by the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). *Eur Heart J*. 2017;38:3382-90.
17. Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield JE, Galie N, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): the Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC). *Eur Heart J*. 2010;31:2915-57.
18. Attenhofer Jost CH, Tan NY, Hassan A, Vargas ER, Hodge DO, Dearani JA, et al. Sudden death in patients with Ebstein anomaly. *Eur Heart J*. 2018;39:1970-7.
19. Fratz S, Janello C, Müller D, Seligmann M, Meierhofer C, Schuster T, et al. The functional right ventricle and tricuspid regurgitation in Ebstein's anomaly. *Int J Cardiol*. 2013;167:258-61.
20. Burri M, Vogt MO, Hörer J, Cleuziou J, Kasnar-Samprec J, Kühn A, et al. Durability of bioprostheses for the tricuspid valve in patients with congenital heart disease. *Eur J Cardiothorac Surg*. 2016;50:988-93.

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