The outcomes of operations for 539 patients with Ebstein anomaly

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Objective: Our objective was to review the long-term outcomes of patients with Ebstein anomaly who underwent cardiac surgery at our institution.

Methods: Patient records were reviewed, and all patients were mailed a medical questionnaire or contacted by means of telephone. Patients who had pulmonary atresia with an intact ventricular septum, complex conotruncal abnormalities, and atrioventricular discordance with ventriculoarterial discordance were excluded.

Results: From April 1, 1972, to January 1, 2006, 539 patients with Ebstein anomaly had 604 cardiac operations. The mean age at the time of the initial operation at our institution was 24 years (range, 8 days–79 years). Three hundred seventeen of the patients were female. One hundred forty-three (26.5%) patients had a prior invasive cardiac procedure before coming to Mayo Clinic. At the time of the first operation at Mayo Clinic, 182 patients had tricuspid valve repair, and 337 had tricuspid valve replacement. The 30-day mortality was 5.9% for the entire cohort (2.7% after 2001). Late survival was 84.7% at 10 years and 71.2% at 20 years. In a multivariate analysis of overall mortality for the patients' first operation at Mayo Clinic, increased hematocrit values, pulmonary valve stenosis, tricuspid valve replacement, absence of ablation of an accessory pathway, miscellaneous arrhythmia procedure, branch pulmonary artery enlargement, need for mechanical support postoperatively, emergency chest opening in the intensive care unit, and absence of sinus rhythm at dismissal were all predictive of mortality. When only preoperative characteristics were included, increased hematocrit values, mitral valve regurgitation requiring surgical intervention, prior cardiac procedure, and moderate-to-severe to severe reduction in right ventricular systolic function were associated with mortality. Preoperative sinus rhythm and an accessory pathway were associated with survival. Patients rated their health as excellent or good (New York Heart Association class I or II) in 83% of surveys returned.

Conclusion: Ebstein anomaly can be surgically treated with low perioperative mortality. Both tricuspid valve repair and tricuspid valve replacement are associated with good long-term survival. Risk factors for poorer outcome included right, and/or left ventricular systolic dysfunction; increased hemoglobin/hematocrit values; male sex; right ventricular outflow tract obstruction; or hypoplastic pulmonary arteries.

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Ebstein anomaly is a spectrum of tricuspid valve (TV) and right ventricular (RV) dysplasia.¹ The TV usually is insufficient but also might be stenotic. Atrial septal defect (ASD) or patent foramen ovale (PFO) occurs in 30% to

Abbreviations and Acronyms

ASD	= atrial septal defect
ICU	= intensive care unit
LV	= left ventricular
MV	= mitral valve
PFO	= patent foramen ovale
RV	= right ventricular
RVOT	= right ventricular outflow tract
TR	= tricuspid regurgitation
TV	= tricuspid valve
TVrpl	= tricuspid valve replacement
TVrpr	= tricuspid valve repair

70% of cases, and ventricular pre-excitation is associated with approximately 15% of cases. Less commonly, left ventricular (LV) dysfunction and ventricular septal defect are observed. Morbidity and mortality are thought to be related to the degree of TV regurgitation or stenosis; the size, thickness, and function of the right ventricle; and the presence or absence of an ASD. Patients with mild forms of Ebstein anomaly can be symptom free throughout their entire lives, but patients with severe forms can die *in utero*.

The purpose of this report is to assess the long-term outcome of operations for Ebstein anomaly. Specifically, we intend to define operative and long-term mortality and the determinants of operative and long-term mortality, to define the determinants of reoperation, and to compare the outcomes of TV repair (TVrpr) and TV replacement (TVrpl).

Materials and Methods**Patients**

From April 1, 1972, to January 1, 2006, 539 patients with Ebstein anomaly had an operation at the Mayo Clinic performed by one of 2 surgeons (GKD, JAD). Excluded from this cohort are patients with pulmonary atresia with intact ventricular septum, complex conotruncal abnormalities, and atrioventricular discordance with ventriculoarterial discordance ("corrected transposition").

Indications for Operation

The indications for operation included 1 or more of the following: symptoms of dyspnea or right-sided heart failure (New York Heart Association class III or IV), progressive exercise intolerance, tachyarrhythmias not controllable with medication or amenable to catheter-based intervention, and significant associated lesions, including ASDs, ventricular septal defects, or pulmonary stenosis. In some patients, progressive cardiomegaly, a cardiothoracic ratio of greater than 0.65, severe cyanosis, and reduced LV function were an indication for operation.

Operative Management

Operative management of patients with Ebstein anomaly has been described previously and consisted of (1) electrophysiologic mapping for localization of accessory conduction pathways in patients with ventricular pre-excitation; (2) closure of any ASD or PFO; (3) elimination of previously placed shunts and correction of any as-

sociated anomalies, such as ventricular septal defect, pulmonary stenosis, or patent ductus arteriosus; (4) performance of any indicated antiarrhythmia procedures, such as surgical division of accessory conduction pathways, cryoablation of atrioventricular nodal re-entry tachycardia, or the right-sided maze procedure; (5) consideration of plication of the atrialized right ventricle; (6) reconstruction of the TV when feasible or valve replacement; and (7) excision of redundant right atrial wall (right reduction atrioplasty).^{1,2} In a few patients a bidirectional cavopulmonary shunt was performed when the right ventricle was markedly dilated and functioning poorly.³

Our initial repair, reported in 1979, consisted of plication of the free wall of the atrialized portion of the right ventricle, posterior tricuspid annuloplasty, and right reduction atrioplasty.⁴ The repair was based on the construction of a monocusp valve by using the anterior leaflet; it brought the functional tricuspid annulus up to the true tricuspid annulus. More recently, we have incorporated various modifications of TVrpr based on the numerous variants of Ebstein anomaly subsequently encountered.¹ This includes TVrpr at the level of the functional tricuspid annulus by bringing the anterior papillary muscle or muscles toward the ventricular septum to facilitate coaptation of the leading edge of the anterior leaflet with the ventricular septum, adding an anterior annuloplasty when indicated, and selective plication or resection of the atrialized RV. In this experience the 2 most important features that enabled a successful durable repair were a free leading edge of the anterior leaflet and at least 50% delamination of the anterior leaflet. TVrpr is performed during aortic crossclamping, and therefore the valve anatomy could be accurately assessed and repair sutures could be placed with minimal motion trauma. After completion of a valve repair, the TV is tested by means of bulb syringe injection of saline into the right ventricle with temporary pulmonary artery occlusion. Before 1982, intraoperative assessment to detect residual tricuspid regurgitation (TR) after valve repair was performed by introducing an exploring finger into the right atrium through the appendage for direct palpation of the TV in the beating heart. Intraoperative assessment of the TV after repair or replacement was performed with epicardial echocardiography from 1982 through 1985, after which transesophageal echocardiography was used routinely.

When the TV could not be reconstructed, the valve tissue adjacent to the RV outflow tract was excised, and a prosthetic valve (bioprosthetic more often than mechanical) was inserted.⁵ The suture line was deviated cephalad to the atrioventricular node, bundle of His, and membranous septum to avoid injury to the conduction system. The suture line was deviated cephalad to the tricuspid annulus posterolaterally, where the tissues were frequently very thin, to avoid injury to the right coronary artery. The struts of the bioprosthetic valve were oriented so that they straddled the membranous septum and conduction tissue. The valve sutures were tied while the heart was perfused and beating to detect any rhythm abnormalities.

At the beginning of our experience, a portion of the atrialized right ventricle was routinely excised. However, postoperative ventricular arrhythmias were common, and we believed this could be related to ventricular suture lines, compromise of small branches of the right coronary artery, or both. In addition, we have a new appreciation that in some patients, the thin-walled atrialized right ventricle maintains contractility. Our current practice is to plicate or resect an atrialized ventricle selectively. This is most often performed when it is thin, transparent, and dyskinetic on echocardiography.

Data Collection

Two of the investigators (MLB and DJD) collected all pertinent data from the Mayo Clinic medical record. This included all follow-up information regarding reoperation and deaths that had been received from non-Mayo Clinic-referring physicians and health care providers.

In addition to review of medical records, vital status was assessed through an online database (www.Accurint.com). Detailed health status questionnaires were mailed to all participants not known to be dead. Patients who did not return or complete the questionnaire were sent a second questionnaire. If the second questionnaire was not returned or completed, attempts were made to contact the patient by means of telephone. Two hundred eighty-five patients (64% of 448 alive patients) completed questionnaires and Health Insurance Portability and Accountability Act forms. Forty-three patients completed a questionnaire but did not complete the Health Insurance Portability and Accountability Act form, and 28 patients refused to complete the survey.

Statistical Analysis

Statistical analysis included the Fisher exact test and the χ^2 test of association for comparing proportions. Logistic regression was carried out to analyze early (≤ 30 days) mortality. Log-rank tests and Cox proportional hazards models were used to identify univariate and multivariate predictors of overall mortality, late (> 30 days) mortality, and late (> 30 days) reoperation. In addition, late survival free from reoperation was determined by using death or reoperation as an event. Kaplan–Meier survival curves were drawn for all late outcomes. Predictor variables were analyzed as continuous or categorical variables, as appropriate. Continuous variables were dichotomized when either statistically or clinically appropriate. Multivariate model building was based on a stepwise selection method. The SAS 9.1.3 statistical software system (SAS, Institute Inc, Cary, NC) was used throughout. Only 2-tailed probability values are reported.

Independent variables included, among others: age at operation; year of operation; surgeon; patient sex; age at diagnosis; cardiothoracic ratio on chest radiographic analysis; presence and type of preoperative arrhythmia; rhythm on electrocardiographic analysis; blood oxygen saturation (oximetry); hemoglobin value; hematocrit value; and previous cardiac procedures (see the complete list and definitions shown in [Appendix E1](#)). Associated cardiac defects and type of operation (including TVrpr, TVrpl, and miscellaneous concomitant procedures) also were included. Echocardiographic variables included preoperative and postoperative RV size and function; degree of tricuspid stenosis, regurgitation, or both; degree of pulmonary stenosis or regurgitation; degree of mitral valve (MV) stenosis or regurgitation; and LV ejection fraction. The degree of TR, RV or LV systolic dysfunction, and RV enlargement was graded as none, mild, moderate, moderate-to-severe, and severe on transthoracic echocardiographic analysis. Echocardiographic variables with more than 15% missing data (and up to 45%) were not included in the stepwise selection for the initial multivariate model. To assess their effects, a second multivariate model was fit by first forcing all the variables chosen for the initial multivariate model and then putting in the variables with missing data one by one to see whether they are significant after adjusting for the effects from the initial multivariate model.

The initial operation at Mayo Clinic was TVrpr for 182 patients, TVrpl for 337 patients, and a variety of nonvalve operations for 20 patients ([Figure 1](#)). Of the patients who initially had TVrpr at the

Figure 1. Operations on the tricuspid valve (TV) at Mayo Clinic.

Mayo Clinic, 1 patient subsequently underwent reoperation, and 35 subsequently had TVrpl ([Figure 1](#)). In both the univariate and multivariate analyses, the 35 patients who had both TVrpr and TVrpl were included both in the repair group and in the replacement group. Also, 6 patients who had "other" operations and subsequent TVrpl were included in the replacement group. Thus the TVrpr group had 182 patients, and the TVrpl group had 378 patients.

Results

Patient Characteristics at Initial Operation at Mayo Clinic

The mean patient age was 24.1 ± 18.1 years (range, 8 days–79 years), with 317 (58.8%) patients being female ([Table E1](#)). There was only 1 newborn in this cohort (≤ 30 days at the time of the operation) and 12 patients under the age of 1 year at time of the first operation at Mayo Clinic. Operations took place between April 1, 1972, and January 1, 2006. Most patients had associated cardiac anomalies ([Tables E2 and E3](#)). The most common was an ASD, which was present in 452 (83.9%) of the patients. An accessory conduction pathway was demonstrated by means of electrophysiologic testing in 74 (13.7%) patients, and a history of pre-excitation was obtained from 91 (16.9%) patients.

One hundred forty-three patients had prior cardiac procedures before coming to the Mayo Clinic ([Table 1](#)). The 4 most common procedures were closure of an ASD or PFO in 41 (7.6%) patients, surgical ablation of an accessory pathway in 35 (6.5%) patients, placement of a systemic to pulmonary artery shunt in 33 (6.1%) patients, and TVrpr or TVrpl in 18 (3.3%) patients. Rereplacement of a tricuspid prosthesis was done for significant prosthesis regurgitation, stenosis, or both. None were replaced for patient–prosthesis mismatch.

Initial Operations at Mayo Clinic

The initial operation at Mayo Clinic was TVrpl for 337 (62.5%) patients and TVrpr for 182 (33.8%; [Table 2](#)) patients. An ASD or PFO was closed in 450 (83.5%) patients.

TABLE 1. Previous cardiovascular procedures performed elsewhere

	Patients	
	Total, n = 539	%
Any prior cardiovascular procedure	143	26.5
Closure of ASD or PFO	41	7.6
Ablation of accessory pathway, catheter or surgical	35	6.5
Systemic-to-pulmonary artery shunt	33	6.1
Miscellaneous prior procedures	29	5.4
Tricuspid valve repair or replacement	18	3.3
Cavopulmonary shunt	12	2.2
Closure of ventricular septal defect	7	1.3
Pulmonary valvotomy	7	1.3
Coronary stenting or CABG	5	0.9
Repair of coarctation	5	0.9
Closure of patent ductus arteriosus	5	0.9
Others	6	1.1

ASD, Atrial septal defect; PFO, patent foramen ovale; CABG, coronary artery bypass grafting.

Overall Morbidity

The most common early complication after the first operation at Mayo Clinic (excluding patients who died ≤30 days postoperatively) was an atrial arrhythmia (atrial fibrillation, flutter, supraventricular tachycardia, or premature atrial contractions), which occurred in 15% of patients (Table E4). Low cardiac output occurred in 5.7%, respiratory insufficiency occurred in 5.5%, and transient third-degree atrioventricular block occurred in 4.5%. Eleven (2.2%) patients had permanent atrioventricular block.

The most common reason for early reoperation (≤30 days after the index operation) was for bleeding (16 patients) and delayed sternal closure (16 patients; Table E5). Eleven patients required emergency reopening of the chest in the intensive care unit (ICU). Permanent pacemakers were inserted in 21 patients: 12 for third-degree atrioventricular block and 9 for miscellaneous reasons.

Entire Cohort

Overall total mortality. Data from all 539 patients were used in this analysis. The 30-day and 1-, 5-, 10-, 15-, and 20-year survivals were 94%, 92%, 88%, 85%, 81%, and 71%, respectively (Figure 2 and Table 3). In a multivariate model pulmonary valve stenosis, a higher hematocrit value, TVrpl, branch pulmonary artery enlargement, emergency opening of the chest in the ICU, a miscellaneous arrhythmia procedure (cryoablation, left-sided maze procedure, or ligation of the left atrial appendage), and postoperative need for mechanical cardiac support were associated with higher mortality. Ablation of an accessory pathway and sinus rhythm at discharge were associated with lower mortality.

When only preoperative variables were included in the model, a higher hematocrit value, MV regurgitation requiring

TABLE 2. Surgical procedures at the time of the first Mayo Clinic operation

	Patients	
	Total, n = 539	%
Closure of ASD or PFO	450	83.5
Right reduction atriotomy	400	74.2
Tricuspid valve replacement	337	62.5
Anterior right pericardiectomy	270	50.1
Plication of atrialized right ventricle	186	34.5
Tricuspid valve repair	182	33.8
Intraoperative ablation of accessory pathway(s)	63	11.6
Right-sided maze procedure	52	9.7
Repair of pulmonary stenosis	33	6.1
Closure of systemic-to-pulmonary artery shunt	26	4.8
Closure of ventricular septal defect	23	4.3
Miscellaneous arrhythmia surgery	19	3.5
RVOT enlargement	15	2.8
Creation of shunt	12	1.7
Ablation of AV nodal re-entrant tachycardia	10	1.9
Mitral valve replacement or repair	10	1.9
Coronary artery bypass grafting	10	1.9
Repair partial atrioventricular septal defect	8	1.5
Branch pulmonary artery enlargement	6	1.1
Pulmonary valve replacement	5	0.9
Closure of patent ductus arteriosus	5	0.9
Fontan with tricuspid valve excision	4	0.7
Mechanical assist device or transplantation	4	0.7

ASD, Atrial septal defect; PFO, patent foramen ovale; RVOT, right ventricular outflow tract; AV, atrioventricular.

surgical repair, and moderate-to-severe and severe RV systolic dysfunction were associated with higher mortality, and an accessory pathway was associated with lower mortality.

There were numerous variables associated with mortality in a univariate fashion (Table 3). Those of particular interest included increased preoperative RV size and decreased RV systolic function, preoperative decreased LV systolic

Figure 2. Long-term survival for all 539 patients. Time 0 is the time of the first cardiac operation at the Mayo Clinic.

TABLE 3. Univariate and multivariate predictors of early mortality (<30 days), late mortality (>30 days), and survival free from late reoperation (>30 days) for all patients at the initial operation at the Mayo Clinic

All patients	Early mortality			Late mortality			Survival free from late reoperation		
	<i>P</i> value, uni	OR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv
Demographics									
Male				.019			.031	1.48	.038
Younger age at operation (<12 y)							.002	3.23	<.001
Younger age at diagnosis (↓ 10 y)							.001		
Preoperative CT ratio >0.65	.033						.019		
Preoperative O ₂ saturation ≤85%*				.035					
Preoperative hemoglobin >15 g/dL	.016			.016					
Preoperative hematocrit (↑ 10%)	.016			.020	1.53	.014			
Echocardiography*									
Moderate-severe to severely enlarged preoperative RV size				.025					
Moderate-severe to severely reduced preoperative RV function	.029	2.895		.002					
Moderate to severely reduced preoperative LV function	.056			<.001			.004	2.31	.019
Moderate-severe to severely enlarged postoperative RV size				.002			.018		
Moderate-severe to severely reduced postoperative RV function	.001			.001			.004	2.44	.001
Moderate to severely reduced postoperative LV function	<.001			.010	3.67	<.001	.025		
PV stenosis, mild or greater							<.001	2.00	.045
Pulmonary artery stenosis/hypoplasia							<.001		
Associated heart disease									
Accessory conduction pathway				.021	0.28	.034			
Coronary artery disease									
MV regurgitation requiring surgical intervention				<.001	8.17	.006	<.001		
Pulmonary artery stenosis or hypoplasia							.034		
Severe PV regurgitation							<.001		
Surgical procedures									
No TV repair				.029			.004		
TV replacement				.008			.009		
No ablation of accessory pathway(s)				.017	0.21	.033			
Repair PV stenosis				.043			.040		
CABG									
Permanent pacing wires	.019								
Miscellaneous arrhythmia procedure				.005			.002	3.75	.008
MV replacement or repair	.019			.025					
RVOT enlargement				.003	4.90	.001	.007		
Branch PA enlargement				.037	4.40	.041			
Pulmonary valve replacement							.008		
LVAD/transplantation/IABP	.020								
Prior cardiac procedures									
Any prior procedure				.048			.036		
Prior pulmonary valvotomy				.020					
Prior systemic-to-pulmonary shunt							.039		
Prior cavopulmonary shunt							<.001	3.07	.006
Early nonfatal complication									
Atrial arrhythmias				.002			.026		
Wide complex tachycardia				.002			.029	2.45	.007
Respiratory insufficiency				.042			<.001		

TABLE 3. Continued

All patients	Early mortality			Late mortality			Survival free from late reoperation		
	<i>P</i> value, uni	OR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv
Renal insufficiency				<.001	6.79	<.001			
Dialysis							.006		
Low cardiac output							.001		
Thrombus in RA or TV							.032		
Deep sternal infection							.022	11.78	<.001
Intra-aortic balloon pump							.007		
Early reoperation									
Tamponade	<.001	15.99	<.001				.044		
LVAD, RVAD, or mechanical support	<.001	82.54	<.001	<.001			<.001		
Creation of shunt	.004			<.001					
Closure of shunt							.026		
Emergency opening of chest in ICU	<.001	31.33	<.001	<.001					
Miscellaneous early reoperation	<.001			.002			.009		
Preoperative arrhythmia and laboratory data									
Past history of any arrhythmia							.013		
No accessory pathway				.027					
Patients in sinus rhythm on preoperative ECG				.003	0.41	.020			
Increased QRS duration				.001	1.15	.001			

uni, Univariate; OR, Odds ratio; mv, multivariate; CT, cardiothoracic; RV, right ventricular; LV, left ventricular; PV, pulmonary valve; MV, mitral valve; TV, tricuspid valve; CABG, coronary artery bypass grafting; RVOT, right ventricular outflow tract; PA, pulmonary artery; LVAD, left ventricular assist device; IABP, intra-aortic balloon pump; RVAD, right ventricular assist device; ICU, intensive care unit; ECG, electrocardiography. *Missing data (>15% and <45%).

function, cardiothoracic ratio of greater than 65% determined by means of chest radiography, and number of variables related to a right ventricular outflow tract (RVOT) obstruction or small pulmonary arteries.

Overall early mortality. There were 33 deaths 30 days or less after the operation. In a multivariate model emergency opening of the chest in the ICU, postoperative need for mechanical cardiac support, and early tamponade were associated with higher mortality. When only preoperative variables were entered into the model, moderate-to-severe and severe RV systolic dysfunction were the only variables associated with higher mortality (Table 3).

Overall late mortality. When the 33 early deaths were excluded, the 1-, 5-, 10-, 15-, and 20-year survivals were 98%, 94%, 90%, 86%, and 76%, respectively. In a multivariate model MV regurgitation requiring operative repair, RV outflow tract enlargement, enlargement of the pulmonary arteries, renal insufficiency, and increased QRS duration were associated with increased mortality. Late survival was associated with ablation of an accessory pathway and preoperative sinus rhythm (Table 3).

When only preoperative variables were entered into the model, MV regurgitation requiring operation, increased hematocrit values, and LV dysfunction were associated with greater mortality and an accessory conduction pathway and the presence of preoperative sinus rhythm were associated with lower mortality.

Overall freedom from late reoperation. The 1-, 5-, 10-, 15-, and 20-year freedoms from any (on the TV or other) reoperation were 97%, 91%, 82%, and 70%, respectively. In a multivariate model hypoplastic or stenotic pulmonary arteries, insertion of an intra-aortic balloon pump postoperatively, prior cavopulmonary shunt, younger age at the time of the operation, and earlier era of operation were associated with late reoperation. When only preoperative variables were entered into the model, pulmonary valve stenosis, hypoplastic or stenotic pulmonary arteries, prior cavopulmonary

Figure 3. Survival free from late reoperation (on the tricuspid valve or other) for all patients after the first cardiac operation at the Mayo Clinic.

shunt, younger age at the time of the operation, and an earlier era of operation were associated with late reoperation.

Survival free from late operation. The 1-, 5-, 10-, 15-, and 20-year survivals and freedoms from any (on the TV or other) reoperation were 95%, 86%, 74%, 62%, and 46%, respectively (Figure 3). In a multivariate model the following variables were associated with an increased risk of death or reoperation: deep sternal wound infection, miscellaneous arrhythmia procedure, wide complex tachycardia, moderate-to-severe or severe postoperative RV systolic dysfunction, moderate-to-severe preoperative LV systolic dysfunction, and age less than 12 years at the time of the operation. When only preoperative variables are considered, a prior cavopulmonary shunt, pulmonary valve stenosis, male sex, moderate-to-severe preoperative LV systolic dysfunction, and age less than 12 years were associated with higher risk of death or late reoperation (Table 3).

Recurring variables. Several variables were statistically significant in a univariate model for 3 of the 4 following subsets: overall mortality, early mortality, late mortality, and survival free from late reoperation (Table 3). The variables were male sex, greater preoperative cardiothoracic ratio, higher preoperative hemoglobin or hematocrit values, reduced preoperative RV and LV systolic function, enlarged postoperative RV size, MV regurgitation requiring surgical intervention, no TVrpr, TVrpl, repair of pulmonary valve stenosis, miscellaneous arrhythmia procedure, RVOT enlargement, and need for mechanical support.

TVrpr Versus TVrpl

The patients whose operations at the Mayo Clinic were TVrprs differed from those whose operations were TVrpls in a number of ways. In these comparisons, patients who had TVrpr followed by TVrpl were only included in the TVrpr group. In general, patients who had valve repair were operated on earlier in the series (mean year of operation, 1989 vs 1996). On preoperative transthoracic echocardiographic analysis, the TV was judged to be not repairable in 32% of the repair group and in 85% in the replacement group. On echocardiographic analysis, anatomic severity of the valve malformation was assessed to be moderately severe and severe in 61% of the repair group and 93% of the replacement group. The replacement group had more prior cardiac procedures than the repair group. All of these differences were statistically significant (Table 4). Postoperatively, 13% of the patients who had valve repair had at least moderate residual TR, whereas no patients who had replacement had moderate residual TR.

TVrpr Subset

Overall mortality. Data from 182 patients who had TVrpr were used in this analysis. The 30-day and 1-, 5-, 10-, 15-, and 20-year survivals were 95%, 93%, 93%, 88%, 87%, and 76%, respectively. In a multivariate model higher preop-

TABLE 4. Differences between the TVrpr and TVrpl patient groups

Variable	% TVrpr	% TVrpl	P value
Anatomic severity (moderate-to-severe/severe)	60.82	93.38	<.001
TV repairable on echocardiography, yes	46.67	7.08	<.001
TV repairable on echocardiography, equivocal	21.33	7.50	
Pulmonary artery architecture, hypoplastic	1.65	6.12	.040
Pulmonary artery architecture, discrete stenosis	0.55	1.46	
Postoperative RV function (grade 3 or 4+)	15.71	48.73	<.001
Postoperative TV regurgitation (grade 3 or 4+)	12.71	0.00	<.001
Any prior cardiac procedure	13.74	34.11	<.001
Prior closure of atrial septal defect	1.65	11.37	<.001
Prior TVrpr	0.00	3.79	.008
Prior systemic-to-pulmonary artery shunt	2.75	7.00	.043
Prior ablation of accessory pathway	2.75	8.75	.009
Right reduction atrioplasty	86.81	76.97	.007
Plication of atrialized right ventricle	69.78	18.95	<.001
Right-sided maze procedure	3.85	13.12	.001
Perioperative atrial arrhythmia, atrial fibrillation, atrial flutter, PACs, SVT	8.24	17.20	.005
Perioperative low cardiac output state	2.20	7.00	.020
Respiratory insufficiency	2.20	6.71	.036
Perioperative thrombus on TV or in the right atrium	0.00	2.62	.031
Postoperative permanent pacemaker, third-degree HB	0.55	3.50	.041
Secondary sternal closure	0.00	4.37	.002
Past history of a specific arrhythmia	47.25	56.27	.049
Inducible ventricular tachycardia on EP study	6.04	1.75	.008
Patients in sinus rhythm preoperatively	96.70	91.84	.031

TVrpr, Tricuspid valve repair; TVrpl, tricuspid valve replacement; TV, tricuspid valve; RV, right ventricular; PACs, premature atrial contractions; SVT, supraventricular tachycardia; HB, heart block; EP, electrophysiologic.

erative hematocrit values and previous cardiac operations were associated with higher mortality. Preoperative sinus rhythm was associated with lower mortality. There were numerous variables associated with mortality in a univariate fashion (Table 5). Those of particular interest included preoperative decreased RV or LV systolic function, lower preoperative blood oxygen saturation, and higher hemoglobin levels.

Early mortality. There were 9 deaths at 30 days or sooner after the operation. There were too few deaths to develop a multivariate model. In a univariate model reduced preoperative RV and LV systolic function, the need for permanent pacing wires postoperatively, and the need for a ventricular assist device postoperatively were associated with mortality (Table 5).

Late mortality. When the 9 early deaths were excluded, the 1-, 5-, 10-, 15-, and 20-year survivals were 98%, 98%,

TABLE 5. Variables significant in a univariate model and multivariate model for tricuspid valve repair

	Early mortality†		Late mortality >30 d		Survival free from late reoperation		
	<i>P</i> value, uni	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv
Demographic							
Male sex		.043			.050	1.83	.043
Preoperative hematocrit >45%		.049					
Echocardiography*							
Moderate-severe or severely reduced preoperative RV function	.035						
Moderate to severe reduction in preoperative LV function	.034	.007			.014		
Moderate to severe MV regurgitation					<.001		
Mild or greater pulmonary valve stenosis					<.001	5.14	.005
Pulmonary artery stenosis or hypoplasia					.008		
Moderate-severe to severely reduced postoperative RV function		.016					
Moderate to severe reduction in postoperative LV function		<.001			.005	7.61	.010
Associated heart disease							
Coronary artery disease		.001			.022		
Subvalvular pulmonary stenosis/RVOT obstruction					.028		
Miscellaneous associated cardiac defects		.010					
Previous CV procedure							
Any prior cardiac procedure		<.001	5.97	<.001	.003	2.33	.020
Prior cavopulmonary shunt		.007			<.001		
Prior closure of ventricular septal defect		.022					
Prior coronary stenting or CABG		.001			.002		
Concomitant procedures							
Right-sided maze procedure					.003	6.12	.021
CABG		.001					
Permanent pacing wires	.002						
Early, nonfatal complications					<.001		
Low cardiac output state							
Early reoperation							
Emergency opening of the chest in ICU	.012						
Preoperative arrhythmia							
Cardiac arrest before surgical intervention		.029			.001		
Patients in sinus rhythm preoperatively		<.001	0.08	<.001			

uni, Univariate; HR, hazard ratio; mv, multivariate; RV, right ventricular; LV, left ventricular; MV, mitral valve; RVOT, right ventricular outflow tract; CV, cardiovascular; CABG, coronary artery bypass grafting; ICU, intensive care unit. *Missing values (>15% and <45%). †No multivariate model because of low number of events.

93%, 92%, and 80%, respectively. In a multivariate model the previous cardiac procedure was associated with increased mortality, and preoperative sinus rhythm was associated with lower mortality (Table 5).

Freedom from late reoperation. The 1-, 5-, 10-, 15-, and 20-year freedoms from reoperation on the TV were 100%, 95%, 89%, 77%, and 64%, respectively. In a multivariate model prior surgical procedure and increased QRS duration were associated with lower freedom from reoperation. Plication of an atrialized right ventricle reduced the need for late reoperation. When only preoperative variables were entered into the model, patients with a prior cardiac surgical procedure were associated with a lower freedom from reoperation.

The freedom from reoperation rate over a 20-year period was 51%, 83%, and 61% for patients 10 years of age or less, 11 to 20 years of age, and more than 20 years of age, respectively ($P = .032$).

Survival free from late reoperation. The 1-, 5-, 10-, 15-, and 20-year survivals free from reoperation on the TV was 98%, 93%, 84%, 73%, and 56%, respectively. In a multivariate model a greater risk of death or reoperation was associated with the following variables: right-sided maze procedure, pulmonary valve stenosis, a previous cardiovascular procedure, greater QRS duration, moderate-to-severe LV systolic dysfunction postoperatively, and male sex. When only preoperative variables were included, male sex, pulmonary valve

stenosis, and previous cardiovascular procedures were associated with greater risk of death or late reoperation (Table 5).

Recurring variables. Several variables were statistically significant in a univariate model for 3 of the 4 following subsets: overall mortality, early mortality, late mortality, and survival free from late reoperation (Table 5). The 4 variables included male sex, reduced preoperative and postoperative LV systolic function, any prior cardiac procedure, prior cavopulmonary shunt, and prior stenting or coronary artery bypass grafting procedure.

TVrpl Subset

Overall mortality. Data from 378 patients who had TVrpl were used in this analysis. Of these 378 patients, 43 (11.4%) had a prior TVrpr. The 30-day and 1-, 5-, 10-, 15-, and 20-year survivals were 94%, 91%, 86%, 83%, 74%, and 68%, respectively. In a multivariate model repair of pulmonary valve stenosis, miscellaneous arrhythmia procedure, mechanical circulatory support postoperatively, and emergency opening of the chest in the ICU were associated with higher mortality. Surgical ablation of an accessory pathway and postoperative sinus rhythm were associated with lower mortality.

When only preoperative variables were included in the model, MV regurgitation requiring an operation, and reduced LV systolic function were associated with increased mortality. The presence of an accessory conduction pathway was associated with lower mortality. There were numerous variables associated with higher mortality in a univariate fashion (Table 6). Those of particular interest included increased preoperative RV size and decreased RV systolic function, preoperative decreased LV systolic function, radiographic cardiothoracic ratio of greater than 0.65, lower blood oxygen saturation, and higher hemoglobin levels.

Early mortality. There were 23 deaths at 30 days or less after the operation. In a multivariate model emergency opening of the chest in the ICU and postoperative need for a ventricular assist device were associated with increased mortality. When only preoperative variables were entered into the model, moderate and severe LV systolic dysfunction was the most significant variable associated with mortality, and after adjusting for it, no other risk factors were significant (Table 6).

Late mortality. When the 23 early deaths were excluded, the 1-, 5-, 10-, 15-, and 20-year survivals were 97%, 92%, 88%, 79%, and 72%. In a multivariate model RVOT enlargement, wide complex tachycardia, early reoperation, and QRS duration were associated with higher mortality. An accessory pathway was associated with lower mortality (Table 6). Considering only preoperative variables, MV regurgitation requiring an operation and reduced RV systolic function preoperatively were associated with late mortality. An accessory pathway was associated with late survival.

Freedom from late reoperation on the TV. The 1-, 5-, 10-, 15-, and 20-year freedoms from reoperation were 99%,

95%, 83%, 70%, and 59%, respectively. In a multivariate model younger age, greater MV regurgitation requiring surgical intervention, postoperative need for an intra-aortic balloon pump, and a catheter ablation procedure were associated with lower freedom from reoperation. When only preoperative variables were entered into the model, younger age at the time of the operation, MV regurgitation requiring an operation, a catheter ablation procedure, and a prior cardiac procedure were associated with lower freedom from reoperation.

Survival free from reoperation on the TV. The 1-, 5-, 10-, 15-, and 20-year freedoms from reoperation were 96%, 87%, 74%, 56%, and 44%, respectively. In a multivariate model the following variables were associated with death or late reoperation on the TV: MV regurgitation requiring an operation, postoperative renal insufficiency, wide complex tachycardia, moderate-to-severe preoperative reduction of LV systolic function, and age less than 12 years at the time of operation. Preoperative variables associated with death or late reoperation on the TV were as follows: MV regurgitation requiring an operation, moderate-to-severe preoperative reduction of LV systolic function, and younger age at the time of the operation (Table 6).

Recurring variables. Several variables were statistically significant in a univariate model for 3 of the 4 following subsets and were associated with a lower rate of reoperation: overall mortality, early mortality, late mortality, and survival free from late reoperation (Table 6). They included increased preoperative cardiothoracic ratio, reduced preoperative RV and LV systolic function, reduced postoperative RV systolic function, MV regurgitation requiring surgical intervention, miscellaneous arrhythmia surgery, postoperative wide complex tachycardia, postoperative mechanical support or miscellaneous early operation.

A summary of all independent predictors of overall mortality, early and late mortality, and survival free from reoperation is shown in Table 7.

TVrpr and TVrpl in Patients Less Than 12 and 12 or More Years of Age

Patients were divided into groups less than 12 and 12 or more years of age (Figure 4) and stratified into patients who had TVrpr or TVrpl to examine survival free from late reoperation. This age cutoff was chosen because an adult-sized TV prosthesis could usually be inserted in patients who were 12 years of age or older. Patients were analyzed in an intent-to-treat manner in that patients who received a TVrpr at the Mayo Clinic followed by a TVrpl were included in the TVrpr group only for this analysis ($n = 35$). There was no difference between those who had TVrpr and TVrpl in survival free from late reoperation for patients 12 years of age and older. However, there was a significant advantage for patients who received a TVrpr ($P < .001$) compared with a TVrpl at less than 12 years of age. For patients less

TABLE 6. Variables significant in a univariate and multivariate model for tricuspid valve replacement

	Early mortality (≤ 30 d)			Late mortality (> 30 d)			Survival free from late reoperation		
	<i>P</i> value, uni	OR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv	<i>P</i> value, uni	HR, mv	<i>P</i> value, mv
Demographics									
Younger age at operation < 12 y							$< .001$	0.33	$< .001$
Age at diagnosis ($\downarrow 10$ y)							.008		
$\uparrow$ Preoperative CT ratio	.003			.01			.045		
Preoperative decreased oxygen saturation*							.011		
Echocardiography*									
Moderate-severe to severely enlarged preoperative RV size				.019					
Moderate-severe to severely decreased preoperative RV function	.033			.003			.021		
Moderate to severely reduced preoperative LV function				.011	2.79	.009	.008	2.28	.005
Moderate-severe to severely enlarged postoperative RV size				.018					
Moderate-severe to severely reduced postoperative RV function	.001	3.08	.028	.038					
Moderate to severely reduced postoperative LV function	.006								
Associated heart disease									
Accessory conduction pathway				.013	0.11	.029			
Mitral regurgitation requiring surgical intervention				.006			$< .001$	7.13	.001
Prior cardiac procedures									
Any prior cardiac procedure	.027								
Prior closure of atrial septal defect	.020								
Concomitant procedures									
Anterior right pericardiectomy				.005					
Ablation of accessory pathway(s)									
Repair PV stenosis				.011					
Systemic-to-pulmonary artery shunt							.015		
Miscellaneous arrhythmia procedure				.006			.041		
Mitral valve replacement or repair				.001	5.34	.025	.017		
RVOT enlargement				$< .001$	8.09	$< .001$			
LVAD/transplantation/IABP	.010								
Early nonfatal complication									
Ventricular arrhythmia, PVCs, ventricular tachycardia, or ventricular fibrillation	.034								
Wide complex tachycardia, ventricular tachycardia or ventricular fibrillation				$< .001$	3.32	.002	.002	2.42	.007
Atrial arrhythmia, atrial fibrillation, atrial flutter, PACs, SVT	.02			.029					
Low cardiac output state							.006		
Thrombus in RA or on TV							.023		
Respiratory insufficiency				.023			$< .001$		
Renal insufficiency				.001	3.33	.028	$< .001$	3.96	.003
Dialysis				.011			$< .001$		
Early reoperation									
Tamponade	$< .001$								
LVAD, RVAD, or mechanical support	$< .001$	125.4	$< .001$	$< .001$			$< .001$		
Creation of shunt									
Pericardiectomy									
Emergency opening of chest in ICU	$< .001$	33.4	$< .001$						
Miscellaneous early reoperation	.002			.034	9.79	.003	$< .001$		
Preoperative arrhythmia data									
Any preoperative arrhythmia							.027		
Accessory pathway				.01					
Patients with pre-excitation on preoperative ECG				.031					

uni, Univariate; OR, odds ratio; mv, multivariate; HR, hazard ratio; CT, cardiothoracic; RV, right ventricular; LV, left ventricular; PV, pulmonary valve; RVOT, right ventricular outflow tract; LVAD, left ventricular assist device; IABP, intra-aortic balloon pump; PACs, premature atrial contractions; SVT, supraventricular tachycardia; RA, right atrium; TV, tricuspid valve; RVAD, right ventricular assist device; ICU, intensive care unit; ECG, electrocardiography. *Missing data ($> 15\%$ and $< 45\%$).

TABLE 7. Summary of hazard ratios of multivariate models

Overall mortality			
	All patients (n = 539)	TV repair (n = 182)	TV replacement (n = 378)
All variables			
LVAD, RVAD, or mechanical support	17.77		32.11
Emergency opening of chest in ICU	9.90		17.85
Enlargement of pulmonary arteries	4.97		
Pulmonary stenosis: mild or greater	3.20		
Miscellaneous arrhythmia procedure	3.18		3.84
Tricuspid valve replacement	1.92		
HCT results (increase by 10 units)	1.75		
Sinus, discharge rhythm by ECG	0.25	0.224	0.38
Ablation of accessory pathways	0.20		0.09
Repair of pulmonary stenosis			3.81
Sinus, preoperative rhythm by ECG		0.17	
Preoperative variables only			
Mitral regurgitation requiring surgical intervention	3.07		5.89
Preoperative RV function moderate-severe to severely reduced	2.76		
HCT results (increase by 10 units)	1.46	1.61	
Accessory pathway, associated cardiac defect	0.51		0.27
Preoperative LV function, moderate to severely reduced			2.75
Previous cardiovascular procedure		3.44	
Sinus, preoperative rhythm by ECG		0.15	
Early mortality			
	All patients	TV repair	TV replacement
All variables			
LVAD, RVAD, or mechanical support	82.54		125.36
Emergency opening of chest in ICU	31.33		33.43
Tamponade	15.99		
Preoperative variables only			
Preoperative RV function, moderate-severe to severely reduced	2.90		
Preoperative LV function, moderate to severely reduced		3.08	
Late mortality			
	All patients	TV repair	TV replacement
All variables			
Enlargement of RV outflow tract	4.90		8.09
Miscellaneous early reoperation			9.79
Branch PA enlargement	4.40		
Prior cardiovascular procedure		5.97	
Renal insufficiency, early postoperative complication	6.79		3.33
Wide complex tachycardia			3.32
MV regurgitation requiring operative treatment	8.17		
QRS duration (increase by 10 units)	1.15		1.14
Sinus, preoperative rhythm by ECG	0.41	0.08	
Accessory pathway on EP study			0.11
Ablation of accessory pathways	0.21		
Preoperative variables only			
Mitral regurgitation requiring surgical intervention	9.86		5.51
Prior cardiovascular procedure		5.97	
Preoperative RV function: moderate-severe to severely reduced			2.79
Sinus, preoperative rhythm by ECG	0.42	0.08	
Accessory pathway	0.28		0.28
Increased HCT (10 units)	1.53		
Preoperative LV function, moderate to severely reduced	2.27		2.79

TABLE 7. Continued

	Survival free from late reoperation		
	All patients	TV repair	TV replacement
All variables			
Male sex		1.29	
Age <12 y	3.23		0.33
Preoperative LV function: moderate to severely reduced	2.31		2.28
Postoperative LV function: moderate to severely reduced		7.61	
Pulmonary stenosis: mild or greater		2.36	
Postoperative RV function: moderate severe to severely reduced	2.44		
Previous cardiovascular procedures		2.14	
Mitral valve regurgitation requiring surgical intervention			7.13
Right-sided maze procedure		8.05	
Miscellaneous arrhythmia procedure	3.75		
Wide complex tachycardia	2.45		2.42
Deep sternal wound infection	11.78		
Renal insufficiency			3.96
Prior cavopulmonary shunt			
Increased QRS duration (↑ 10 S)		1.13	
Preoperative variables only			
Male sex	1.48	1.93	
Age <12 y	2.63		2.78
Preoperative LV function: moderate to severely reduced	2.27		2.44
Pulmonary valve stenosis: mild or greater	2.00	5.37	
Previous surgical procedure		2.22	
Prior cavopulmonary shunt	3.07		
Mitral valve regurgitation requiring surgical intervention			11.37

TV, Tricuspid valve; LVAD, left ventricular assist device; RVAD, right ventricular assist device; ICU, intensive care unit; HCT, hematocrit; ECG, electrocardiography; RV, right ventricular; LV, left ventricular; PA, pulmonary artery; MV, mitral valve; EP, electrophysiologic; HR, hazard ratio.

than 12 years of age, those who had a TVrpl were statistically more likely to have the following characteristics: younger age at the time of diagnosis, more recent date of operation, reduced LV systolic function, more anatomically severe Ebstein anomaly, more likely to be “unrepairable” by means of echocardiographic analysis, have hypoplastic or stenotic pulmonary arteries, have an enlarged right ventricle postoperatively, and less likely to have a plication or resection of an atrialized portion of the ventricle.

Predictive Value of Preoperative Echocardiography

The echocardiographer attempted to predict the likelihood of successful surgical valve repair in 284 cases. The sensitivity was 59%, the specificity was 92%, the positive predictive value was 65%, and the negative predictive value was 90%.

Protective Effect of Pre-excitation

In several analyses the presence of ventricular pre-excitation or its likely surrogates (ie, preoperative arrhythmia, ablation of accessory pathways) was associated with improved survival. The only differences between patients with and without pre-excitation were surgeon (surrogate for surgical division of bypass tract), degree of MV regurgitation, prior closure of an ASD or PFO, history of an

arrhythmia, year of operation, and age at the time of the operation. There did not appear to be a relationship between the severity of the Ebstein anomaly and the presence of pre-excitation.

Discussion

The management of Ebstein anomaly requires a choice between nonoperative treatment and operative interventions. This choice may be difficult because of the very broad spectrum of severity of Ebstein anomaly. Patients with very severe forms of Ebstein anomaly can die *in utero* or shortly after birth, but patients with mild forms of the anomaly can be completely asymptomatic for their entire life and never need specific treatment. There are no randomized trials comparing the results of surgical treatment with those of nonsurgical treatment. Celermajer et al⁶ found that greater severity of Ebstein anomaly, cardiothoracic ratio of greater than 60%, RVOT obstruction, and presentation as a fetus were associated with poorer survival. Interestingly, ventricular pre-excitation and era of diagnosis were not associated with increased risk of death. Attie et al⁷ also reported that poorer outcome was associated with more severe forms of Ebstein anomaly, male sex, cyanosis, worse functional status, severe TR, and early age at diagnosis in 72 unoperated adult

Figure 4. A and B, Survival free from late reoperation on the tricuspid valve (TV) for patients less than 12 or 12 or more years of age. The 35 patients who had TV replacement and had prior TV repair followed by TV replacement were removed from the TV replacement group.

patients. Thus the indications for operation we used in the current study seem reasonable.

Our study is consistent with the findings of previous studies^{6,7} because male sex, RVOT obstruction, cyanosis, and more severe forms of the anomaly were associated with poorer outcome. Cardiomegaly was associated with poorer outcome in our study in a univariate, but not in a multivariate, model.

Celermajer et al⁶ showed no effect of pre-excitation on outcome, and in our study ventricular pre-excitation was associated with better outcome. Many patients in this study were operated on before the era of transvenous catheter ablation. We speculated that the improved survival occurred because the primary indication for operation was for surgical division of the bypass tract, and these patients might have had less severe forms of Ebstein anomaly at the time they had their operations.^{9,10} However, except for the fact that patients who had pre-excitation were younger than those without pre-excitation, our data did not support this speculation. It might also be that patients who have but are unknown to have a bypass tract might die later because of a ventricular

arrhythmia. Thus patients who have a bypass tract and receive proper treatment might have a survival advantage when compared with patients who have not undergone adequate treatment.

Echocardiographic Assessment of Valve Reparability in Ebstein Anomaly

Favorable echocardiographic criteria for TVrpr include both the valve leaflet location and morphology and the papillary muscle location and attachments. Valves that have severe leaflet displacement into the RV apex or those anteriorly rotated into the RVOT are generally not suitable for the traditional monocusp repair. These forms of Ebstein anomaly leave very little effective right ventricle below the point of coaptation. If the echocardiographer does not view TV tissue in the apical 4-chamber view, the valve will typically need to be replaced. However, it is possible that the apical 4-chamber view can be angled anteriorly, so that some of the outflow tract is in view. In this projection one might falsely believe that the septal leaflet is present near the true TV inlet. The internal cardiac crux should be identified to avoid this pitfall. Views from the parasternal short-axis projection at the base of the heart can aid in identifying the proximity of TV tissue in the RVOT to the pulmonary valve. If the anterior leaflet is tethered at multiple sites (most importantly the tip), then the surgeon might have difficulty mobilizing the leaflet, and coaptation with the septum will be ineffective. The apical 4-chamber projection can be used to determine the tethering sites of the anterior leaflet. Positioning of the papillary muscles can be assessed from most imaging planes and is important for assessment of leaflet mobility. Dense papillary muscle attachments to the anterior leaflet can prohibit a traditional repair. Sacrificing these attachments might result in important residual TV regurgitation after repair.

Valves that have multiple leaflet fenestrations (hence multiple jets of TR) might be difficult to repair because of a deficiency of useable tissue. Color Doppler assessment of TR can be difficult because RV systolic pressure is usually normal in these patients, and regurgitant jets can appear as very laminar color flow. In addition, these jets can be oriented in directions that are not usually expected. The severity of TR is frequently underestimated by the inexperienced echocardiographer. Principles of color Doppler scanning are based on jet velocity and not on the quantity of regurgitation. This can have important implications because surgical intervention might be inappropriately deferred if the amount of TR is underestimated. The preoperative echocardiographic assessment of the patient with Ebstein anomaly requires meticulous use of all imaging planes and experience gathered from evaluating large numbers of these patients.

Caveats

In this study we hoped to identify the determinants of poor outcome and the determinants of good outcome. Intuitively,

and based on prior small studies, one might suggest that RV size and function, as well as LV systolic function and other measures of severity of TV dysplasia, would correlate with outcome. Because our study is retrospective, we had to use echocardiographic data obtained over a 33-year period. These data were not reported in a uniform fashion. Also, over the study, measurement of RV size and function was (and remains) imperfect by means of echocardiographic analysis. In addition, because of leftward displacement of the ventricular septum, it is difficult to know the implications of measured LV ejection fraction. Despite these caveats, RV size and both RV and LV systolic function are important determinants of outcome. Improved measurements of ventricular size and function, such as could be obtained with magnetic resonance imaging, might increase the usefulness of ventricular size and function in predicting outcome.

Mortality

Previously, we reported early mortality of 6.3% for 189 patients who underwent TVrpr or TVrpl (these 189 patients are an early part of the current cohort). In the current era mortality ranges from 2.5% to 31%.^{2,5,8,11-21} In a collaborative study of 150 patients, the early mortality was reported as 13%.⁸ In the current report the early mortality was 6% for all 539 patients, 5% for the 182 patients who had TVrpr, and 6% for the 378 patients who had TVrpl. Considering only the operations since 2001, perioperative mortality decreased to 2.7% (operations = 110).

Total mortality in the current study (20 year; both early and late) was 29% for all patients, 24% for patients who had TVrpr, and 32% for patients who had TVrpl. To our knowledge, there are no other studies of this magnitude and length of follow-up with which to compare our results.

Reoperation

We presented risk of late reoperation in 2 formats: freedom from late reoperation and survival free from late reoperation. In both analyses, 10- and 20-year freedom from reoperation was better in the valve repair than in the valve replacement group. The highest rate of reoperation was in patients less than 12 years of age who had valve replacement. These results, however, are not just a function of whether the valve was replaced or repaired but, more importantly, on the severity of the disease that led to the decision to repair or replace the valve. The decision to repair or replace the valve is guided by whether the valve is amenable to repair with minimal residual valve regurgitation, as well as comorbid conditions, which might sway the surgeon to do the quickest operation with the shortest ischemic time. For example, a patient with severely reduced RV systolic function, moderately reduced LV systolic dysfunction, hypoplastic pulmonary arteries, and secondary erythrocytosis might be better served by valve replacement than by an attempted valve repair associated with moderately-severe residual regurgitation.

Valve Repair Versus Valve Replacement

An important question is whether it is better to repair or replace the TV. Intuitively, one might suggest that the operation associated with lower mortality and lower reoperation rate would be the preferable operation. However, if the goal of the operation is to reduce or eliminate the volume overload of the right atrium and ventricle, then the operation that is associated with a lesser degree of residual TR might be preferable. In this study the 20-year survival for TVrpr was 76%, and that for TVrpl was 67%. Survival free from reoperation at 20 years for valve repair was 53% for patients operated on at less than 12 years of age and 57% for patients operated on at more than 12 years of age. Survival free from reoperation at 20 years for valve replacement was 16% for patients operated on at less than 12 years of age and 60% for patients operated on at more than 12 years of age.

One must resist the temptation to conclude that valve repair is inherently superior to valve replacement based on these data. It is clear that those patients who had valve replacement had more severe disease than those who had valve repair. In addition, significant residual TV regurgitation persisted in 33% of patients who had valve repair but in only 1% of patients who had valve replacement. This would favor valve replacement. It also is important not to conclude that one operation is superior to the other based on short-term outcome because medium- to late-term failure of valve repair clearly occurs.

For patients 12 years of age and older, we believe that the valve should be repaired if there is not more than moderate residual TR at the end of the operation. If there is greater than moderate residual TR, we believe the valve should be replaced because of the long-term effects of TR on RV function. For patients younger than 12 years of age, greater degrees of TV regurgitation might be acceptable because of the relatively greater need for reoperation for repeat valve replacement in this age group.

Numerous techniques have been described for repair of the TV since the first report of Hunter and Lillihai.^{1,2,13,17,20-32} This is not surprising because no 2 hearts with Ebstein anomaly have exactly the same anatomy. In addition, the enormous number of repair techniques is likely the result of incomplete satisfaction with current repair methods already described. Although many of the techniques available report good early results, very few document late outcomes. Undoubtedly, the techniques for valve repair will continue to evolve, and newer promising methods of repair need to be evaluated to determine whether a greater percentage of valves can be repaired while maintaining good long-term durability.^{16,33}

Summary

We believe this is the largest surgical series of patients with Ebstein anomaly. Because we incorporated greater than 125 potential independent variables and assessed numerous

dependent outcomes, there are numerous variables that were related to outcome both in a univariate fashion, multivariate fashion, or both. All of these can be important when trying to predict the early and late outcome of an operation. Table 7 provides a summary of the variables that were statistically significantly associated with 1 or more of the dependent outcome variables in a multivariate model.

In conclusion, it appears that male sex, RVOT obstruction (including pulmonary stenosis and hypoplastic or stenotic pulmonary arteries), MV regurgitation requiring surgical intervention, cyanosis, and of the RV and/or LV systolic dysfunction are among the more important preoperative variables that are predictive of higher mortality. These variables should be helpful in stratifying patients' potential operative risk.

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Discussion

Dr Vaughan A. Starnes (Los Angeles, Calif). I would like to congratulate Dr Dearani on an excellent and clear presentation. Dr Dearani and his colleagues have presented a comprehensive review of the Mayo Clinic's experience with surgical treatment of 539 patients with Ebstein anomaly over a 34-year period of time. I believe this represents the largest and most comprehensive review thus far. The Mayo operation consisted of TVrpl in 61%, TVrpr in 34%, concomitant ASD closure in 84% and plication of the right ventricle in 35%, and ablation of the pathway in 13%. The early mortality was excellent at a low value of 6%, and the overall survival of the entire cohort treated at 10 and 20 years was 85% and 71%, respectively. An extensive analysis of variables predicting early and late mortality

were analyzed. As we have seen in the presentation, a multivariate analysis was performed, and predictors of overall mortality were mitral regurgitation, moderate-to-severe RV dysfunction, RVOT or pulmonary artery reconstruction, and TVRpl.

This leads me to my first question, Joe, which is one of 3.

In your paper and presentation TVRpr is advocated and seems to be related to better survival, and yet it seems you have replaced more valves than you repaired, and this tendency seems to be increasing in the more recent experience. Based on this large experience, what are the determinants currently used to see whether the valve is repairable, and what degree of insufficiency would you accept?

Dr Dearani. Thank you for your remarks, Dr Starnes. We prefer TVRpr when the valve is repairable. In recent years, there has been a tendency for more replacement, which we believe is the result of 2 major reasons. First, in the adult population the durability of a tricuspid bioprosthesis is excellent, and the freedom from reoperation is actually much better with a bioprosthesis than with a fair repair. The second reason is that recently there has been a greater percentage of patients who have already undergone a previous operation elsewhere, and in our experience the chances to rerepair a valve in that setting are rarely successful.

Dr Starnes. My second question—and I think I know your bias but to clarify it—if the valve is to be replaced, which valve are we going to use, a mechanical or a tissue valve?

Dr Dearani. Of approximately 350 TVRpls, about 300 were bioprosthetic, and 45 were mechanical. We prefer bioprostheses for 2 reasons: first, the durability is excellent, and second, our ability to successfully treat atrial arrhythmias with the concomitant maze procedure has improved in recent years. Consequently, we are able to optimize the ability to spare the patient from long-term warfarin anticoagulation.

The patients we would consider for mechanical TVRpl would really be those who require anticoagulation for another reason, specifically a left-sided mechanical prosthesis, which occurs very infrequently. There are some important observations that we have made with regard to mechanical TVRpl in Ebstein anomaly that are worth mentioning. First, it is easy to insert a very large prosthesis because the true tricuspid annulus is usually severely dilated. Caution should be exercised that the prosthesis is not oversized when a mechanical valve is being used. In the presence of RV dysfunction, which is always present in Ebstein anomaly, the discs might not open and close normally. Thus the risk of thrombosis-related problems with a mechanical prosthesis is greater in this setting, and therefore there is a need for even stronger anticoagulation. For all of these reasons, we prefer a porcine bioprosthesis when valve replacement is required.

Dr Starnes. The last question, Joe, and maybe the most difficult to answer is this: Given the variability of a clinical presentation of Ebstein anomaly and your extensive experience, when should a patient undergo an operation?

Dr Dearani. The timing of the operation can be challenging for some patients. In general, clinical deterioration and progressive symptomatology with Ebstein anomaly occurs over a long period of time, generally years. Consequently, it is uncommon to advise an operation at the initial outpatient visit. When symptoms are absent, the decision to operate is usually based on serial changes that occur on echocardiograms. We have learned from this study that the presence of significant RV dysfunction is a powerful predic-

tor of outcome. With this in mind, an operation should be advised before significant RV dysfunction, and I will say that that is not always easy to ascertain. Determination of RV function is challenging. We currently use both echocardiographic analysis and magnetic resonance imaging to quantitate the function of the right ventricle, and on many occasions we have found marked discrepancies in the results between these 2 diagnostic modalities. We continue to learn from both echocardiography and magnetic resonance imaging, but at the current time, there are limitations for both.

Other indications for surgical intervention include abnormal exercise tolerance, cyanosis, the presence or progression of arrhythmias, and progressive increase in RV size. If echocardiography suggests high probability of TVRpr, then we would advise an earlier operation.

Dr Starnes. Again, I would like to congratulate the author on an excellent presentation and the manuscript that was given to me in advance, and I would like to thank the Association for the floor.

Dr Afksendiyos Kalangos (*Geneva, Switzerland*). How do you evaluate RV function, by means of echocardiography or magnetic resonance imaging, and what are the criteria for RV dysfunction of more than moderate degree?

Dr Dearani. As I indicated earlier, echocardiography is very imperfect. In my experience echocardiography can tell you whether the RV function is good or whether it is bad; everything in between is very difficult to quantitate. Magnetic resonance imaging evaluation appears to be more objective in terms of being able to provide an ejection fraction. However, in a given patient there have been circumstances when the echocardiography suggested good RV function and the magnetic resonance imaging suggested poor RV function and vice versa.

Therefore there is a lot to be learned in terms of quantitating RV function in Ebstein anomaly. I do not have the answer. It is very imperfect.

Dr Antonio Corno (*Liverpool, United Kingdom*). I enjoyed the presentation. You presented a very small number of patients with one and a half type of ventricular repair, and you say these have been done in the last few years. Actually, you are limiting the indication to one and a half ventricular repair to the patient with severe RV dysfunction. Do you consider that if you largely extend the indication to one and a half ventricular repair, as done in other series, you can dramatically reduce the number of TVRpls? If you use the one and a half ventricular type of repair, you reduce one third of the systemic venous return through the TV, and this is allowing you to do a much better TVRpr in terms of reduced degree of residual stenosis or regurgitation, and this would allow you to reduce the need for TVRpl.

Dr Dearani. I agree. The point that is being made is whether the addition of a bidirectional Glenn shunt improves the percentage of successful repairs, and I think the answer is, in general, yes. With that in mind, the bidirectional Glenn shunt is not a perfect solution either. One issue with Ebstein anomaly that we are going to investigate in the future is exercise tolerance in patients who have had a bi-ventricular repair with TVRpl versus those patients who have had a bidirectional Glenn shunt (ie, a one and a half ventricle repair).

The one and a half ventricle repair, in my experience, helps reduce the early mortality in patients with profound RV dysfunction, but the late results with the bidirectional Glenn shunt are unclear. In addition, in this patient subgroup there is a high probability of late

arrhythmias, and the bidirectional Glenn shunt does affect the ability to access the heart in the standard fashion for electrophysiologic evaluation. Reducing the volume and size of the right ventricle with a bidirectional Glenn shunt might enable a greater number of successful tricuspid repairs by decreasing tension on suture lines and allowing for a smaller annulus size without creating tricuspid stenosis. At the current time, we apply the bidirectional Glenn shunt selectively.

Dr Pedro Becker (*Santiago, Chile*). Do you have information on whether the RV dysfunction gets better after surgical intervention? Did you evaluate that and whether it is related to prognosis? Also, what has been your incidence of heartbeat block in TVrpl?

Dr Dearani. In our experience the incidence of heart block is 2% or less with TVrpl. We place the suture line cephalad to the true tricuspid annulus and conduction tissue.

Investigation of preoperative and postoperative RV function is another area of great interest. Most investigation is being done with magnetic resonance imaging evaluation. Ebstein anomaly is an RV myopathy, and all of the maneuvers that we do in the operating room do not take away the fact that a myopathy is still present. We believe that plication or resection of the atrialized right ventricle is reasonable and should be done at the discretion of the surgeon. We believe that reducing the size of a markedly dilated right ventricle might increase the efficiency of the right ventricle and perhaps indirectly improve function. We know that the size of the right ventricle, in general, gets smaller after you eliminate the regurgitant lesion. It is not well documented at this time whether this results in improvement in RV function. Magnetic resonance imaging examination preoperatively and postoperatively should help delineate this in the future.

Appendix E1. Independent variables of interest

Demographics and laboratory data

Age at operation
 Age at diagnosis by history
 Patient sex
 Surgeon (1 and 2)
 Year of operation
 Preoperative cardiothoracic ratio on chest radiography
 Preoperative oxygen saturation
 Preoperative hemoglobin
 Preoperative hematocrit

Echocardiography

Preoperative right ventricular size
 Preoperative right ventricular systolic function
 Preoperative tricuspid valve regurgitation severity
 Preoperative left ventricular systolic function
 Anatomic severity of Ebstein anomaly
 TV repairable: yes, no, equivocal
 Mitral valve systolic regurgitation
 Pulmonary valve stenosis
 Architecture of pulmonary arteries: hypoplastic, stenotic, or normal
 Postoperative right ventricular size
 Postoperative right ventricular systolic function
 Postoperative tricuspid valve regurgitation severity
 Postoperative left ventricular systolic function

Associated heart disease

Atrial septal defect or patent foramen ovale
 Accessory conduction pathway
 Pulmonary valve stenosis
 Ventricular septal defect
 Atrioventricular node re-entry tachycardia
 Bilateral superior venae cavae
 Patent ductus arteriosus
 Partial atrioventricular septal defect
 Coronary artery disease on angiography
 Anomalous pulmonary venous connection
 Pericarditis
 Hypertrophic obstructive cardiomyopathy
 Absent coronary sinus
 Mitral valve regurgitation requiring surgical intervention
 Subvalvular pulmonary stenosis/right ventricular outflow tract obstruction
 Pulmonary artery stenosis or hypoplasia
 Pulmonary valve regurgitation
 Miscellaneous associated cardiac defects not included in any above

Previous cardiovascular procedure

Any prior cardiac procedure, including surgical procedures, pacemakers, and catheter-based interventions
 Prior closure of atrial septal defect
 Prior tricuspid valve repair
 Prior tricuspid valve replacement
 Prior systemic-to-pulmonary artery shunt
 Prior cavopulmonary shunt
 Prior permanent pacemaker
 Prior ablation of accessory pathway: catheter based and surgical based

Prior closure of ventricular septal defect
 Prior pulmonary valvotomy
 Prior tricuspid valve replacement
 Prior pulmonary artery banding
 Prior coronary stenting or coronary artery bypass grafting
 Prior repair of coarctation
 Prior closure of patent ductus arteriosus
 Prior enlargement of right ventricular outflow tract
 Prior complete pericardiectomy
 Prior left superior vena cava ligation

Surgical procedure

Closure of atrial septal defect
 Right reduction atrioplasty
 Plication of atrialized right ventricle
 Anterior right pericardiectomy
 Tricuspid valve repair
 Tricuspid valve replacement
 Ablation of accessory pathway(s)
 Right-sided maze procedure
 Closure of any shunt
 Systemic-to-pulmonary shunt
 Cavopulmonary shunt
 Repair of pulmonary valve stenosis
 Closure of ventricular septal defect
 Ablation of atrioventricular node re-entrant tachycardia
 Fontan procedure
 Coronary stenting or coronary artery bypass grafting
 Repair of coarctation
 Repair of partial atrioventricular septal defect
 Systemic-to-pulmonary artery shunt
 Coronary artery bypass grafting
 Repair of partial anomalous pulmonary veins
 Permanent pacing wires
 Miscellaneous arrhythmia procedure: ablation of right or left isthmus or left-sided maze procedure
 Mitral valve replacement or repair
 Complete pericardiectomy
 Pulmonary valve replacement
 Myectomy
 Enlargement of right ventricular outflow tract
 Branch pulmonary artery enlargement
 Electrophysiologic mapping
 Closure of patent ductus arteriosus
 Mechanical support and transplantation
 Miscellaneous operation not listed above

Early nonfatal complications (<30 d)

Third-degree heart block, transient (<3 d)
 Third-degree heart block, permanent
 Ventricular arrhythmia: premature ventricular contractions, ventricular tachycardia, or fibrillation
 Wide complex tachycardia: ventricular tachycardia or fibrillation
 Junctional tachycardia
 Atrial arrhythmia: atrial fibrillation, atrial flutter, premature atrial contractions, supraventricular tachycardia
 Miscellaneous arrhythmia not listed above
 Myocardial infarction
 Cerebrovascular accident

Low cardiac output state: inotropes >3 d	LVAD, RVAD, or mechanical support
Intra-aortic balloon pump inserted in the intensive care unit	Creation of shunt
Respiratory insufficiency: ventilated >3 d	Pericardiectomy
Renal insufficiency: >3 mg/dL	Closure of shunt
Dialysis	Emergency opening of the chest in the intensive care unit
Gastrointestinal bleed	Miscellaneous early reoperation not listed above
Deep sternal wound infection	Arrhythmia data
Extracorporeal membrane oxygenation	Past history of any arrhythmia (including palpitations and other arrhythmias)
Sepsis or endocarditis or pneumonia	Past history of a specific arrhythmia (excluding palpitations)
Neurologic complication: seizures or transient ischemic attack	Cardiac arrest before operation (before entering the operating theater)
Thrombus on tricuspid valve or in right atrium requiring anticoagulation	Accessory pathway on electrophysiologic study
Postpericardial syndrome	Atrioventricular nodal re-entry tachycardia on electrophysiologic study
Hematologic complication	Inducible ventricular tachycardia on electrophysiologic study
Miscellaneous complication not otherwise specified	Inducible ventricular fibrillation on electrophysiologic study
Early reoperation	Catheter ablation procedure on electrophysiologic study
Bleeding	Patients in sinus rhythm preoperatively on electrocardiography
Tamponade	Patients with pre-excitation on preoperative electrocardiography
Permanent pacemaker for third-degree heart block	Patients in sinus rhythm at dismissal on electrocardiography
Permanent pacemaker (miscellaneous reason)	QRS duration on dismissal or follow-up electrocardiography
Tracheostomy	
Delayed sternal closure	
Wound debridement	
Tricuspid valve reoperation	

TABLE E1. Patient age at date of operation

Age at operation (y)	All patients (first operation at Mayo Clinic; n = 539)		Tricuspid valve repair group (n = 182)		Tricuspid valve replacement group (n = 378)	
	N	%	N	%	N	%
≤10	156	29	39	21	112	30
11-20	115	21	51	28	68	18
>20	268	50	92	51	198	52

TABLE E2. Associated cardiac defects

	Patients	
	Total (n = 539)	%
ASD or PFO	452	83.9
Ventricular septal defect	23	4.3
Pulmonary stenosis requiring surgical intervention	32	5.9
Subvalvular pulmonary stenosis/RVOT obstruction	9	1.7
Pulmonary artery stenosis or hypoplasia	8	1.5
Severe pulmonary regurgitation	4	0.7
Coronary artery disease by catheterization	9	1.7
Partial atrioventricular septal defect	8	1.5
Bilateral superior venae cavae	7	1.3
Patent ductus arteriosus	6	1.1
Absent coronary sinus	7	1.3
Mitral regurgitation (moderate to severe)	7	1.3
Pericarditis	5	0.9
Anomalous pulmonary venous connection	4	0.7
Hypertrophic obstructive cardiomyopathy	4	0.7
Miscellaneous cardiac defects	45	8.4

ASD, Atrial septal defect; PFO, patent foramen ovale; RVOT, right ventricular outflow tract.

TABLE E3. Associated electrophysiologic abnormalities

	Patients	
	Total (n = 539)	%
Accessory conduction pathway	74	13.7
Atrioventricular nodal re-entry tachycardia	18	3.3
Inducible ventricular tachycardia or fibrillation	18	3.3
Cardiac arrest before surgical admission	15	2.8

TABLE E4. Early complications (patients who died within 30 days are excluded)

	Patients	
	Total (n = 506)	%
Atrial arrhythmia: atrial fibrillation, atrial flutter, PACs, SVT	77	15.2
Ventricular arrhythmia: PVCs, Ventricular tachycardia, or ventricular fibrillation	72	14.2
Wide complex tachycardia: ventricular tachycardia or ventricular fibrillation only	42	8.3
Low cardiac output state	29	5.7
Respiratory insufficiency	28	5.5
Transient third-degree heart block	23	4.5
Junctional tachycardia	21	4.1
Sepsis or endocarditis or pneumonia	18	3.6
Renal insufficiency	12	2.4
Permanent third-degree heart block	11	2.2
Thrombus on tricuspid valve or in right atrium	9	1.8
Intra-aortic balloon pump	8	1.6
Dialysis	8	1.6
Deep sternal wound infection	6	1.2
Miscellaneous arrhythmias	5	1.0
Neurological complication: seizures, TIA	5	1.0
Hematologic complication	3	0.6
Myocardial infarction	2	0.4
Cerebrovascular accident	1	0.2
Gastrointestinal bleed	1	0.2

PACs, Premature atrial contractions; SVT, supraventricular tachycardia; TIA, transient ischemic attack.

TABLE E5. Early cardiac reoperations (within 30 days)

	Patients	
	Total (n = 539)	%
Bleeding	16	3.0
Secondary sternal closure	16	3.0
Miscellaneous other early reoperations	13	2.4
Permanent pacemaker for third-degree HB	12	2.2
Emergency opening of the chest in the intensive care unit	11	2.0
Tamponade	10	1.9
Permanent pacemaker for miscellaneous reason	9	1.7
LVAD, RVAD, or mechanical support in perioperative period	6	1.1
Tricuspid valve reoperation	4	0.7
Wound debridement	4	0.7
Closure of shunt	3	0.6
Pericardiectomy	3	0.6
Creation of shunt	2	0.4

HB, Heart block; LVAD, left ventricular assist device; RVAD, right ventricular assist device.