Background. The purpose of this study was to examine early and late outcome of tricuspid valve repair for Ebstein’s anomaly in young children.

Methods. Between October 1974 and November 2003, 52 children (25 boys) underwent tricuspid valve repair and annuloplasty for Ebstein’s anomaly. Mean age was 7.1 ± 3.9 years (range, 5 months to 12 years). Concomitant procedures included atrial septal defect closure (n = 46), division of accessory conduction pathways (n = 4), ventricular septal defect closure (n = 3), and other (n = 7).

Results. Early mortality was 5.8% (3 of 52 patients; no mortality since 1984, n = 31). Risk factors were age younger than 2.5 years (p = 0.03) and weight less than 10.7 kg (p = 0.03). Morbidity included transient atrial (n = 11) and ventricular arrhythmias (n = 5), and early reoperation in 3 patients. There was no need for a permanent pacemaker. Mean follow-up was 12.2 ± 7.4 years (maximum, 24.3 years). Actuarial survival at 5, 10, and 15 years was 92.3% ± 3.7%, 89.9% ± 4.3%, and 89.9% ± 4.3%, respectively. Freedom from all reoperations at 5, 10, and 15 years was 91.0% ± 4.3%, 76.9% ± 6.8%, and 61.4% ± 8.8%, respectively. Moderate (grade II) or more tricuspid regurgitation on diastolic echocardiogram was the only risk factor for reoperation (p = 0.04). Tricuspid stenosis did not occur in any patient. At late follow-up, 89% of patients were in New York Heart Association class I or II.

Conclusions. Ebstein’s anomaly in young children can now be repaired with low mortality and good tricuspid valve durability. Tricuspid regurgitation at the completion of operation should be mild or less to minimize need for reoperation. Tricuspid valve repair and annuloplasty did not result in stenosis despite somatic growth. Most patients enjoy an excellent quality of life.

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Tricuspid Valve Repair for Ebstein’s Anomaly in Young Children: A 30-Year Experience

Umar S. Boston, MD, Joseph A. Dearani, MD, Patrick W. O’Leary, MD, David J. Driscoll, MD, and Gordon K. Danielson, MD

Divisions of Cardiovascular Surgery and Pediatric Cardiology, Mayo Clinic and Foundation, Rochester, Minnesota

Ebstein’s anomaly (EA) is a rare cardiac anomaly that is seen in 1 in 210,000 live births [1]. The primary underlying abnormality is failure of adequate delamination of the tricuspid valve (TV) leaflets from the endocardium of the right ventricle. This results in the features that are characteristic of EA, which include apical displacement of the leaflets, particularly the septal leaflet, atrialization of portions of the right ventricle, tricuspid regurgitation (TR), and right ventricular dysfunction. Depending on the degree of delamination and dysplasia of the TV, there are infinite degrees of anatomic severity that can be expressed. Age of patients at clinical presentation can also be quite variable depending on the anatomic severity of the anomaly.

There have been a number of studies evaluating the surgical management of EA in adults because this is the age group in which clinical presentation is most common. To date there are few studies evaluating surgical management of young children with EA. Tricuspid valve repair in children with EA is of particular interest from the standpoint of feasibility and durability of repair and long-term outcome in a group of patients who undergo somatic growth. The aim of this study was to evaluate a 30-year experience with our techniques for repairing the TV in young children with EA.

Patients and Methods

Between October 1974 and November 2003, 186 children age 12 years or younger underwent first-time operation for EA at our institution. All patients had atrioventricular and ventriculoarterial concordance and two ventricles. Of these, 52 who underwent TV repair and annuloplasty form the cohort for this study. There were 117 patients who underwent TV replacement. In the remaining patients, isolated atrial septal defect closure was performed in 7 patients, combined atrial septal defect and ventricular septal defect closure in 5 patients, a bidirectional Glenn anastomosis in 2 patients, Fontan operation in 2 patients, and a modified Blalock-Taussig shunt in 1 patient. Patient characteristics are summarized in Table 1. Age of patients at operation ranged from 5 months to 12 years (median, 7.4 years). There were 4 infants, 8 children ages 1 through 2 years, and 40 children ages 3 through 12 years. Weight ranged from 5.7 to 59.1 kg (median, 22.2 kg). The diagnosis of EA was based on clinical evaluation and on echocardiography, cardiac catheterization, or both. Cardiac catheterization was per-
formed more frequently in the early part of the series; this is now performed infrequently and usually only when additional electrophysiologic information is required. Echocardiography can establish the diagnosis using the criterion of apical displacement of the septal leaflet at the crux of the heart by 8 mm/m² or greater [2].

Associated cardiac anomalies are shown in Table 2. Fifty-one of the 52 patients had at least one associated cardiac anomaly.

**Definitions**

Low cardiac output was defined as the need for inotropic support to maintain an adequate blood pressure to sustain organ perfusion. Respiratory insufficiency was defined as ventilator dependence for more than 3 days. Renal insufficiency was defined as a rise in creatinine to greater than 3 g/dL or requiring dialysis. Subjective echocardiographic quantitation of TR after valve repair was from grade 0 to 4: 0 = none, 1 = mild, 2 = moderate, 3 = moderate-severe, and 4 = severe. Tricuspid regurgitation was assessed by using the width of the regurgitant jet.

**Operative Technique**

Our techniques of TV repair for EA have expanded and evolved during the last 30 years [3–7]. Operation is performed by means of a median sternotomy, the ascending aorta is cannulated, and bicaval venous drainage is established. Patients with accessory conduction pathways undergo electrophysiologic mapping for localization followed by surgical division or cryoablation of the pathways [8, 9]. A right-sided maze procedure is performed in patients with a history of intermittent or chronic atrial flutter or atrial fibrillation [9–11]. Components of the operation that have remained constant during the years include establishing a competent TV repair, based primarily on the anterior leaflet of the TV, and tricuspid annuloplasty. Right ventricular plication or limited resection is now performed selectively and usually for thinned out, dysfunctional atrialized right ventricle. A patent foramen ovale is closed by direct suture, and larger atrial septal defects are closed with an autologous pericardial patch. Right reduction atriotomy is routinely performed for an enlarged right atrium. The original technique of TV repair brought the functional tricuspid annulus up toward the true tricuspid annulus; it also effectively plicated the atrialized right ventricle. More recently, our approach often centers on repair of the TV at the level of the functional tricuspid annulus. Ventricular plication or excision is then performed selectively. Details of these techniques are published elsewhere [3–7].

Since the late 1980s, transesophageal echocardiography has become an essential adjunct in the operating room for performing TV repair in patients with EA. Transesophageal echocardiography is performed in all patients after induction of general anesthesia with particular attention to the anterior leaflet, identifying size, mobility, tethering of the leading edge, and degree of fenestrations. In addition, left and right ventricular function, as well as other intracardiac anomalies, is evaluated. Transesophageal echocardiography evaluation is repeated after the patient is weaned from cardiopulmonary bypass to evaluate the competency of the TV repair as well as ventricular function and to exclude residual intracardiac shunts.

**Statistical Analysis**

Demographic and other patient-related data were obtained from Mayo Clinic medical records. Follow-up information was obtained from subsequent clinic visits, written correspondence from local physicians, and mailed questionnaires to patients or families. The rank-sum test was used to compare continuous variables. The probabilities of survival and survivorship free of reoperation were estimated by the Kaplan-Meier method. The associations of potential risk factors for reoperation were assessed with log-rank tests and the Cox proportional hazards model. Data were expressed as mean ± standard deviation, and statistical significance was considered at p less than 0.05. Early operative mortality was defined as death occurring within 30 days of operation or at any time during the index hospitalization. The Mayo Found-

### Table 1. Preoperative Patient Characteristics (52 Patients)

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male/female</td>
<td>25/27</td>
</tr>
<tr>
<td>Mean age at operation</td>
<td>7.1 ± 3.9 years</td>
</tr>
<tr>
<td>Mean weight</td>
<td>23.8 ± 12.7 kg</td>
</tr>
<tr>
<td>Mean body surface area</td>
<td>0.9 ± 0.3 m²</td>
</tr>
<tr>
<td>Mean hemoglobin</td>
<td>16.3 ± 2.9 g/dL</td>
</tr>
<tr>
<td>Mean cardiothoracic ratio</td>
<td>0.7 ± 0.1</td>
</tr>
</tbody>
</table>

**Symptoms**

- Exercise intolerance: 31
- Progressive cyanosis: 25
- Right heart failure: 14
- Progressive cardiomegaly: 13
- Atrial tachyarrhythmias: 11
- Paradoxical emboli: 1

**Previous operations**

- Blalock-Taussig shunt: 3
- BCPS: 1
- Pulmonary valvotomy: 1

*a Some patients had more than one symptom.

BCPS = bidirectional cavopulmonary shunt.

### Table 2. Associated Cardiac Anomalies (51 Patients)

<table>
<thead>
<tr>
<th>Cardiac Anomaly</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>46</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>3</td>
</tr>
<tr>
<td>Hypoplastic pulmonary artery</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary valvular stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Partial atrioventricular canal</td>
<td>1</td>
</tr>
</tbody>
</table>

*a Some patients had more than one anomaly.
Table 3. Concomitant Procedures Performed at Time of Tricuspid Valve Repair

<table>
<thead>
<tr>
<th>Concomitant Procedure</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect closure</td>
<td>46</td>
</tr>
<tr>
<td>Division of accessory pathway</td>
<td>4</td>
</tr>
<tr>
<td>Ventricular septal defect closure</td>
<td>3</td>
</tr>
<tr>
<td>Blalock-Taussig shunt ligation</td>
<td>3</td>
</tr>
<tr>
<td>Pulmonary artery dilatation</td>
<td>2</td>
</tr>
<tr>
<td>Dilatation of SVC-right atrial junction</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary valve replacement</td>
<td>1</td>
</tr>
</tbody>
</table>

* Some patients had more than one concomitant procedure performed at the time of tricuspid valve repair.

SVC = superior vena cava.

Results

Early Results

Concomitant procedures are shown in Table 3. The mean cardiopulmonary bypass time and aortic cross-clamp time were 89.9 ± 25.5 minutes and 50.8 ± 20.5 minutes, respectively.

Preoperative echocardiographic data were available for 33 patients. Mean left ventricular ejection fraction (n = 33), and mean apical displacement of the septal leaflet (n = 12) were 0.572 and 17 mm/m², respectively. As shown in Table 4, 52% of patients had severe TR, and an additional 39% had either moderate or moderate-severe TR.

There were 3 (5.8%) early deaths. Two patients died of persistent low cardiac output despite inotropic support, and 1 patient died of refractory ventricular fibrillation. Since 1984 there have been 31 consecutive TV repairs for EA without mortality in this age group of patients. Univariate analysis demonstrated age younger than 2.5 years (p = 0.03), weight less than 10.7 kg (p = 0.03), and body surface area less than 0.5m² (p = 0.03) to be significant risk factors for early mortality.

Early cardiac morbidity included atrial tachyarrhythmias (n = 11), nonfatal low cardiac output (n = 7), nonsustained ventricular tachyarrhythmia (n = 5), transient complete heart block (n = 4), and right atrial thrombus managed with heparin (n = 2). No patient required a permanent pacemaker. Early pulmonary morbidity included respiratory insufficiency requiring mechanical ventilatory support (n = 3), persistent pleural effusion requiring prolonged chest tube drainage (n = 3), atelectasis secondary to mucous plugs (n = 3; 2 required bronchoscopy), and bronchospasms relieved with steroid treatment (n = 1). Other early morbidity included postdismissal cerebrovascular accident without lasting sequelae (n = 1), lidocaine seizure (n = 1), and renal failure requiring dialysis (n = 1).

Three patients underwent early reoperations for cardiac tamponade (n = 1), mediastinal bleeding (n = 1), and failed repair requiring TV replacement with a porcine bioprosthesis (n = 1).

Table 4 shows the grade of TR in the 33 patients in which transthoracic echocardiograms were obtained at the time of hospital dismissal. The majority of patients (94%) had a TR grade of 2 or less. Mean hospital stay was 9.9 ± 6.8 days.

Late Results

Clinical follow-up was obtained in 98% of early survivors. The mean follow-up (n = 48) was 12.2 ± 7.4 years (range, 10 months to 24.3 years). There were 3 late deaths. One patient who had a massively dilated right ventricle died suddenly 43 days after operation; there were no reported arrhythmias in the postoperative period. Another patient who had a history of atrial tachyarrhythmias died 15.5 years after operation. Both of these patients had right ventricular plications performed during TV repair. The third patient died while awaiting heart transplantation for progressive right ventricular dysfunction and heart failure 6 years after operation as an infant.

Late reoperations and catheter procedures were required in 17 patients (Table 5); no patient had more than one late operation. There were no deaths related to these procedures. Freedom from reoperation from all causes was 91.0% ± 4.3%, 76.9% ± 6.8%, and 61.4% ± 8.8%, at 5, 10, and, 15 years, respectively (Fig 1). There were 13 patients who underwent reoperation for progressive TR; 11 received a bioprosthesis, 1 received a mechanical prosthesis, and 1 had re-repair of the TV. Of these 13 patients who had reoperations, 9 had discharge echocar-

Table 4. Grade of Tricuspid Valve Regurgitation

<table>
<thead>
<tr>
<th>Grade</th>
<th>Preoperative (n = 33)</th>
<th>Postoperative Dismissal (n = 33)</th>
<th>Late Follow-up (n = 31)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>%</td>
<td>n</td>
</tr>
<tr>
<td>0</td>
<td>0</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td>1</td>
<td>3</td>
<td>9</td>
<td>17</td>
</tr>
<tr>
<td>2</td>
<td>8</td>
<td>24</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>15</td>
<td>2</td>
</tr>
<tr>
<td>4</td>
<td>17</td>
<td>52</td>
<td>0</td>
</tr>
</tbody>
</table>

* Preoperative refers to preoperative echocardiogram; postoperative dismissal refers to postoperative echocardiogram at dismissal; and late follow-up refers to echocardiogram at late follow-up in patients who continue to function with tricuspid valve repairs.
diograms at the time of initial repair; 5 of the 9 had a TR grade greater than 2. The mean duration between initial repair and reoperation for TR was 8.8 ± 5.5 years (range, 2.8 to 16.7 years). Freedom from reoperation for TR was 91.0%, 79.6%, and 68.0% at 5, 10, and 15 years, respectively (Fig 2). Univariate analysis demonstrated a TR grade greater than 2 at the time of hospital dismissal to be a significant risk factor for late reoperation ($p = 0.04$). There were no late reoperations for tricuspid stenosis despite somatic growth in all patients. No patient experienced TV endocarditis.

There were 7 patients who underwent late interventional or operative procedures for arrhythmias (Table 5). Three patients had a concomitant right-sided maze procedure performed at reoperation (TV replacement [n = 2] or re-repair [n = 1]). One patient had a surgically placed permanent pacemaker as an isolated procedure, and 1 had permanent pacemaker leads placed prophylactically at the time of TV replacement with a mechanical prosthesis. There were 2 patients who underwent catheter-based radiofrequency ablation to treat paroxysmal ventricular tachycardia.

Late follow-up echocardiographic data were obtained for 94% (31 of 33 patients) of known late survivors who continue to function with TV repairs (Table 4). Seventy-four percent (23 of 31 patients) of patients had a TR grade of 2 or less at late follow-up. There was no patient with tricuspid stenosis.

At last follow-up, New York Heart Association functional class of the 45 known late survivors was I in 28 (62%), II in 12 (27%), III in 4 (9%), and IV in 1 (2%; Fig 3). Survival estimates were 92.3% ± 3.7%, 89.9% ± 4.3%, and 89.3% ± 4.3% at 5, 10, and 15 years, respectively (Fig 4). Univariate analysis, which included ventricular plication, operation before 1985, cardiothoracic ratio greater than 50%, and a TR grade of 2 or greater, found no risk factors for late mortality.

**Comment**

It has not yet been shown whether TV repair or replacement has the better long-term outcome in pediatric patients with EA. Valve replacement exposes the patient to the potential problems of prosthetic valve dysfunction, thromboembolism, endocarditis, and patient-prosthesis mismatch as a result of somatic growth of the patient. It is known that porcine bioprostheses are less durable in

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**Table 5. Late Reoperations and Procedures (17 Patients)**

<table>
<thead>
<tr>
<th>Operation/Procedure</th>
<th>Indication</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>TVR with bioprosthesis</td>
<td>TR</td>
<td>11</td>
</tr>
<tr>
<td>TVR with mechanical valve</td>
<td>TR</td>
<td>1</td>
</tr>
<tr>
<td>TV re-repair</td>
<td>TR</td>
<td>1</td>
</tr>
<tr>
<td>Right-sided maze</td>
<td>AFI</td>
<td>3</td>
</tr>
<tr>
<td>PAPM, leads</td>
<td>AT</td>
<td>1</td>
</tr>
<tr>
<td>Placement PPM leads</td>
<td>P</td>
<td>1</td>
</tr>
<tr>
<td>Pericardiectomy</td>
<td>CRP</td>
<td>1</td>
</tr>
<tr>
<td>Catheter RF ablation</td>
<td>PVT</td>
<td>2</td>
</tr>
</tbody>
</table>

*a Some patients had concomitant late procedures.*

AFI = atrial flutter; AT = atrial tachycardia; CRP = chronic relapsing pericarditis; P = prophylaxis; PAPM, = permanent antitachycardia pacemaker placed intraoperatively; PPM, = permanent pacemaker leads placed intraoperatively; PVT = paroxysmal ventricular tachycardia; RF = radio frequency; TR = tricuspid regurgitation; TV = tricuspid valve; TVR = tricuspid valve replacement.

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Fig 1. Freedom from late reoperation for all causes.

Fig 2. Freedom from late reoperation for recurrent tricuspid valve regurgitation.

Fig 3. New York Heart Association (NYHA) functional class in 45 known late survivors.
In general, we believe that TV repair is preferable to valve replacement whenever repair is feasible. However, if there is failure of delamination of more than 50% of the anterior leaflet or if the leading edge of the leaflet has hyphenated or linear attachment to the right ventricle, a repair may not be obtainable; valve replacement is then preferred. As our experience now shows that valve repair and valve replacement have similar freedoms from reoperation for recurrent TV problems at 10 years, we tend to be more liberal with bioprosthetic valve replacement as opposed to leaving moderate to severe TR after valve repair.

It is recognized that EA is a combination of valvular and ventricular disease, and the right ventricle has both anatomic and functional abnormalities [14–16]. Accordingly, we prefer to send the patient home with either a very good to excellent repair or a bioprosthesis, to eliminate the detrimental effects of ongoing TR on already compromised right ventricular function. Additionally, this current study shows that moderate to severe TR is a significant risk factor for reoperation.

The best treatment for the neonate born with EA is still debated. Twenty to 40% of severely symptomatic neonates with EA will not survive 1 month [17]. Historically, operative mortality for such patients has been prohibitive, but recent improvements in neonatal management and surgery have yielded encouraging results in our institution and elsewhere [18]. Biventricular repairs combined with correction of all associated cardiac defects are feasible, and mid-term results are good [18].

There are controversies regarding the merit of routinely adding a bidirectional cavopulmonary shunt after repair of EA in symptomatic pediatric patients. Disadvantages include longer operating time, loss of catheter access to the right heart from the upper extremities for procedures such as rhythm assessment and ablation or placement of transvenous pacemaker leads, and late sequelae in some patients who are troubled by pulsations in their neck veins. We occasionally use a bidirectional cavopulmonary shunt selectively when the right ventricle is functioning poorly and there is difficulty with weaning the patient from cardiopulmonary bypass. Because concomitant left ventricular dysfunction may be present when the right ventricle fails, it is important to document by direct pressure measurements that the left atrial and pulmonary arterial pressures are low; otherwise the shunt will not be feasible.

We conclude that EA in young children can now be repaired with low mortality and good long-term durability. At the completion of operation, TR should be mild or absent to minimize the need for late reoperation. Tricuspid valve repair and annuloplasty did not result in tricuspid stenosis in spite of somatic growth in all patients. Late survivorship was excellent, and 89% of late survivors were in New York Heart Association functional class I or II.

We thank the Mayo Clinic Division of Biostatistics for statistical support; Massimo Padalino, MD, for early data collection; Judy K. Lenoch for assistance with data collection and analysis; and Evon Heimer for excellent secretarial support.

References


DISCUSSION

DR EZZELDIN A. MOSTAFA (Cairo, Egypt): Actually, all these patients were from the type I and type II classification, as I understood.

The incidence of bidirectional Glenn, you haven’t been confronted by that, as is stated, bidirectional Glenn as a completion for the repair?

DR BOSTON: There were no patients in this cohort who underwent a bidirectional Glenn shunt. We use a bidirectional Glenn procedure selectively, in patients who have severe right ventricular dysfunction and difficulty separating from bypass. In order for the bidirectional Glenn shunt to be a potential adjunct to the operation, it is important for there to be good left ventricular function and low left atrial pressure, which is not always the case in advanced cases of Ebstein’s anomaly.

DR MOSTAFA: My second comment is about the maze. It has been done in the reoperation. Have you been confronted by a maze for the first?

DR BOSTON: We apply a right-sided maze procedure in patients who have atrial fibrillation and/or flutter preoperatively. In this review, there were few children who had atrial arrhythmias preoperatively. Those patients who had preoperative atrial tachyarrhythmias occurred early in the series, prior to the concept of the maze procedure. In the current era, the application of the right-sided or bilateral maze procedure is a routine part of our repair when atrial arrhythmias are present.

DR CHARLES D. FRASER (Houston, TX): Thank you for that very nicely presented series and for confirming what I think we all believe is true, and that is, children who get beyond the neonatal period with this disease can be successfully repaired.

But notably absent from your series are newborns. And I was wondering if you might hypothesize about newborns with Ebstein’s that present with either anatomic or functional pulmonary atresia, do these methods apply to them and what is your suggestion on how to deal with those babies?

DR BOSTON: Our experience with newborns with Ebstein’s anomaly is limited. The youngest infant undergoing repair or replacement of the tricuspid valve was 2 months of age. We are aware of the various approaches for management: single ventricle–Fontan strategy introduced by Starnes and the biventricular approach supported by Knott-Craig. In approximately half of the patients, tricuspid repair was performed. Given the variable results with these different approaches in infants with Ebstein’s anomaly, we believe an individualized approach should be applied, based on the surgeon’s and center’s experience and preference.

DR RALPH S. MOSCA (New York, NY): What he’s pointing out, I believe, is the fact that this technique is a little bit different than what Dr Danielson has been recommending for a while; that is, there is no horizontal plication involved. If that is correct, do you now feel that this part of the repair is no longer necessary?

DR BOSTON: We currently have a selective approach to plication or resection of the atrialized right ventricle. Plication or resection is performed when there is a thinned out, transparent atrialized right ventricle, usually the inferior wall. We do not believe that this type of ventricle contributes to right ventricular function. Our current technique of repair usually involves tricuspid valve repair at the level of the functional annulus, which is the level that the hinge point of the valve exists within the ventricle, with selective plication of the atrialized right ventricle. This is a modification of the original technique described by Dr Danielson in which the atrialized ventricle was plicated routinely as part of the repair.

DR RALPH S. MOSCA (New York, NY): What he’s pointing out, I believe, is the fact that this technique is a little bit different than what Dr Danielson has been recommending for a while; that is, there is no horizontal plication involved. If that is correct, do you now feel that this part of the repair is no longer necessary?

DR BOSTON: The original plication technique was very effective in decreasing the size of the heart and in establishing a durable valve repair. However, in our early years of experience, ventricular arrhythmias were very common. We believed this was due in part to suture lines in the plicated ventricle and/or interruption of small branches of the right coronary artery. We currently plicate or resect the atrialized right ventricle selec-
Since this change in the mid-1980s, the incidence of perioperative ventricular arrhythmias has essentially disappeared. We reserve plication or resection for very thinned, transparent, scarred portions of the right ventricle. In addition, our current repair technique is typically performed at the level of the functional tricuspid annulus. This is in contrast to the original repair where the functional annulus was brought up to the true annulus, which by definition results in plication of the right ventricle.

DR PEDRO J. DEL NIDO (Boston, MA): Two questions. Frequently the source of regurgitation when you’re trying to do a repair is at the point where the anterior leaflet doesn’t quite meet the septum on the leftward medial side. I noticed that you didn’t include in any of your diagrams what sometimes is done, which is dividing the secondary chordae that sometimes tether the anterior leaflet. Do you look at that during your procedure or are those patients automatically excluded because of your criteria, in other words, having a free edge of the anterior leaflet?

DR BOSTON: We believe the two most important anatomical issues that determine successful, durable repair to be first, mobility of the anterior leaflet, and second, the status of the leading edge. Valves amenable to a successful repair usually have at least 50% of the anterior leaflet delaminated and a leading edge that is not adherent to the endocardium. Our experience has shown that maneuvers to detach the leading edge with or without artificial chordae do not provide a satisfactory result.

DR DEL NIDO: You assess that once the heart is open, not by echocardiography?

DR BOSTON: Transthoracic or transesophageal echocardiography is very good at determining the degree of delamination of the tricuspid leaflets. However, assessment of the leading edge of the anterior leaflet can be very difficult even for the experienced echocardiographer. The status of the leading edge of the tricuspid leaflets, especially the anterior leaflet, is best determined at the time of surgery.

DR DEL NIDO: And my second question is a majority of these patients underwent replacement of the valve. Your data show that if you have more than 2+ tricuspid regurgitation, you’re likely to have an early recurrence or require early reoperation. When do you decide to bail out and replace the valve, is it if you see 2+ regurgitation, is that your threshold?

DR BOSTON: The repair is assessed by intraoperative TEE (transesophageal echocardiography) after the discontinuation of bypass. In children, we accept a result of grade II regurgitation or less. We recognize that grade II regurgitation is a risk factor for reoperation; however, the mean time to reoperation was approximately 9 years. We believe this is preferable to valve replacement under most circumstances. In contrast, we resume bypass for an attempt at tricuspid re-repair or replacement if there is grade III or IV regurgitation. Because the myocardium in Ebstein’s anomaly is myopathic, we believe it is important to eliminate the majority of the regurgitation so that right ventricular remodeling with preservation or improvement of RV (right ventricular) function can be optimized.