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# Surgical Management of Congenital (Non-Ebstein) Tricuspid Valve Regurgitation

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**Congenital tricuspid valve regurgitation (TR) is a relatively uncommon condition that includes a heterogeneous group of lesions with a unique management strategy. There are wide anatomic variations that lead to congenital TR in patients without Ebstein malformation. Possible etiologies may include primary valve abnormalities (eg, congenital absence of chordae) or other forms of tricuspid valve dysplasia as in congenitally unguarded tricuspid valve, and patients with pulmonary atresia and intact ventricular septum, which can be similar to Ebstein's valves or secondary regurgitation in association with other anomalies as in atrioventricular septal defects, right ventricular outflow tract obstructive lesions (pulmonary stenosis or atresia with ventricular septal defect [VSD]), tricuspid valve annular dilatation in association with right ventricular volume overload lesions as in congenital coronary arterial fistula with secondary right ventricular enlargement, and Uhl's anomaly. Iatrogenic etiologies in the congenital population include TR secondary to previous VSD closure (chordal or leaflet injury), pacemaker or internal cardiac defibrillator lead-induced TR, and traumatic TR (ruptured chordae). Presentation depends on the severity of the disease and may be apparent in infancy, childhood, or adulthood.**
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## Anatomic and Surgical Considerations

The tricuspid valve has a complex anatomic structure composed of three well-developed leaflets: chordae tendinae, papillary muscles, and the tricuspid annulus. The right atrium and right ventricle (RV) play an important role in that complex structure as well (Fig. 1).[1-9] TV competency depends on successful interaction between these components.

The 3-dimensional complex structure of the TV annulus differs from that of the "saddle-shaped" mitral annulus. It is also a dynamic structure that changes its shape in relation to the cardiac cycle (Fig. 2A-B).[10] All these facts need to be considered during TV repair.[11]

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### Tricuspid Valve Dysplasia

In general, there are two categories of TV dysplasia (Table 1); those with and those without downward displacement. Cases with downward displacement, which is caused by failure of delamination of valve leaflets from the underlying myocardium, are by definition, Ebstein malformation (EM). Although downward displacement is often given as the sole feature of EM, dysplasia of the leaflets and subvalvar apparatus are an almost universal feature as well. The pathognomic findings of EM are downward displacement caused by failure of delamination, whether or not features of dysplasia are present, and a myopathy of the RV that is also a result of failure of delamination. When downward displacement is absent, then the anatomic entity is referred to as "tricuspid valvular dysplasia."[12]

Echocardiography confirms the diagnosis, determines the degree of tricuspid valve regurgitation (TR), allows accurate evaluation of the tricuspid leaflets and subvalvar apparatus (displacement, tethering, dysplasia, etc.), size and function of the right and left ventricles, and detection of septal defects. Magnetic resonance imaging (MRI) is being increasingly used in all types of patients with cardiac disease, including those with EM and other forms of congenital TR. Functional assessment can be made including quantitative measurements of left and right ventricular size and function (Fig. 3). At pres-


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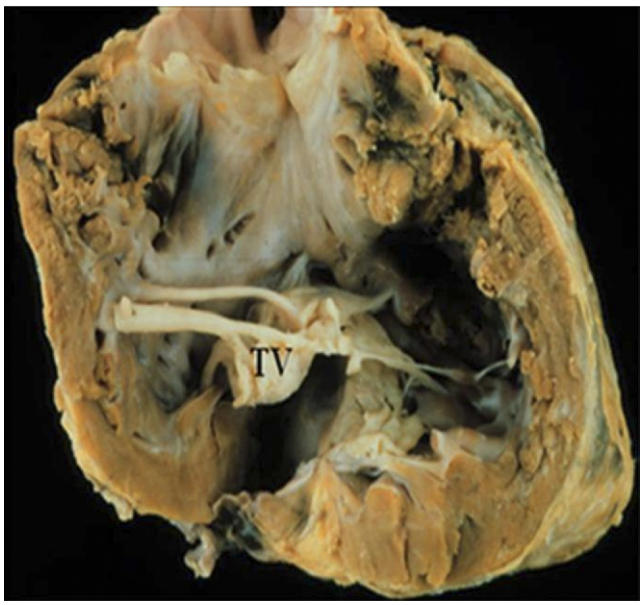


**Figure 1** Pathologic specimens showing tricuspid valve with dysplastic leaflets but no displacement. TV, tricuspid valve. (Reprinted with permission from the Mayo Clinic Foundation for Medical Education and Research.).

**Table 1** Etiologies of Congenital Tricuspid Regurgitation

| |
|---|
| **Primary valvar abnormalities** |
| With Displacement = Ebstein's anomaly |
| Without Displacement = Non-Ebstein's (Tricuspid Valve Dysplasia) |
| TV dysplasia in pulmonary atresia with intact septum (PA/IVS) |
| Congenitally unguarded TV |
| Uhl's anomaly |
| **Secondary valvar abnormalities** |
| *Atrioventricular septal defect (AVSD)* |
| *Right ventricular outflow tract obstruction (RVOTO)* |
| Tetralogy of Fallot (TOF) |
| Pulmonary atresia with ventricular septal defect (PA/VSD) |
| Pulmonary stenosis |
| *Tricuspid regurgitation (TR) with VSD* |
| Gerbode defect |
| *TV annular dilatation (Volume overload lesion)* |
| Coronary artery fistula |
| Atrial septal defect |
| Partial anomalous pulmonary venous connection (PAPVC) |
| *Iatrogenic TR* |
| Detachment of TV leaflet during VSD closure |
| Pacemaker/Cardioverter-defibrillator |

ent, we use echocardiography (2- and 3-dimensional) for evaluation of TV anatomy and MRI for assessment of right and left ventricular size and function.

### TR in the Setting of Pulmonary Atresia with Intact Interventricular Septum

Patients with pulmonary atresia and intact ventricular septum (PA/IVS) tend to have a higher incidence of a regurgitant TV. The mechanism of this regurgitation is different from patients with tetralogy of Fallot (TOF). In TOF, TR is usually caused by right ventricular dilatation with secondary annular dilatation from long-standing pulmonary regurgitation. The degree of TR reflects the degree of right ventricular dilation in these cases, and the progression of TR has been an indication to proceed with pulmonary valve replacement (PVR). In PA/IVS, the TV is usually dysplastic (Fig. 4) and may require surgical intervention from the beginning. In addition, reoperation for persistent or recurrent TR is almost inevitable in this group of patients, and the likelihood of a successful repair and re-repair will decrease with time. The application of various valvuloplasty techniques that have been learned with other TV abnormalities (eg, cone technique for EM) has been helpful in optimizing successful tricuspid valvuloplasty. The addition of a bidirectional cavopulmonary anastomosis (BDCPA) (ie, 1.5 ventricle repair) to the tricuspid valvuloplasty can be considered when there is hypoplasia of the RV or significant RV dysfunction to avoid or delay valve replacement. A single-ventricle pathway can be consid-

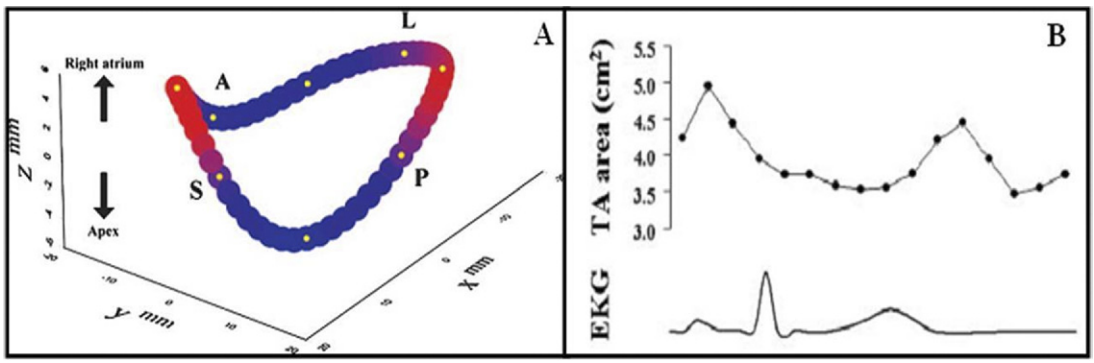


**Figure 2** (A) The reconstructed ring shape for tricuspid annuloplasty based on the average results obtained in healthy subjects at the time of minimum tricuspid annulus area. The positive *x-y-z* axis indicates the respective directions toward the septum, the posterior wall, and the right atrium. The average of each of the manually selected TA locations is indicated by the white dot. A, anterior; L, lateral; P, posterior; S, septum; TA, tricuspid annulus. (B) Dynamic changes in the tricuspid valve annulus during cardiac cycle. (Reprinted with permission.)

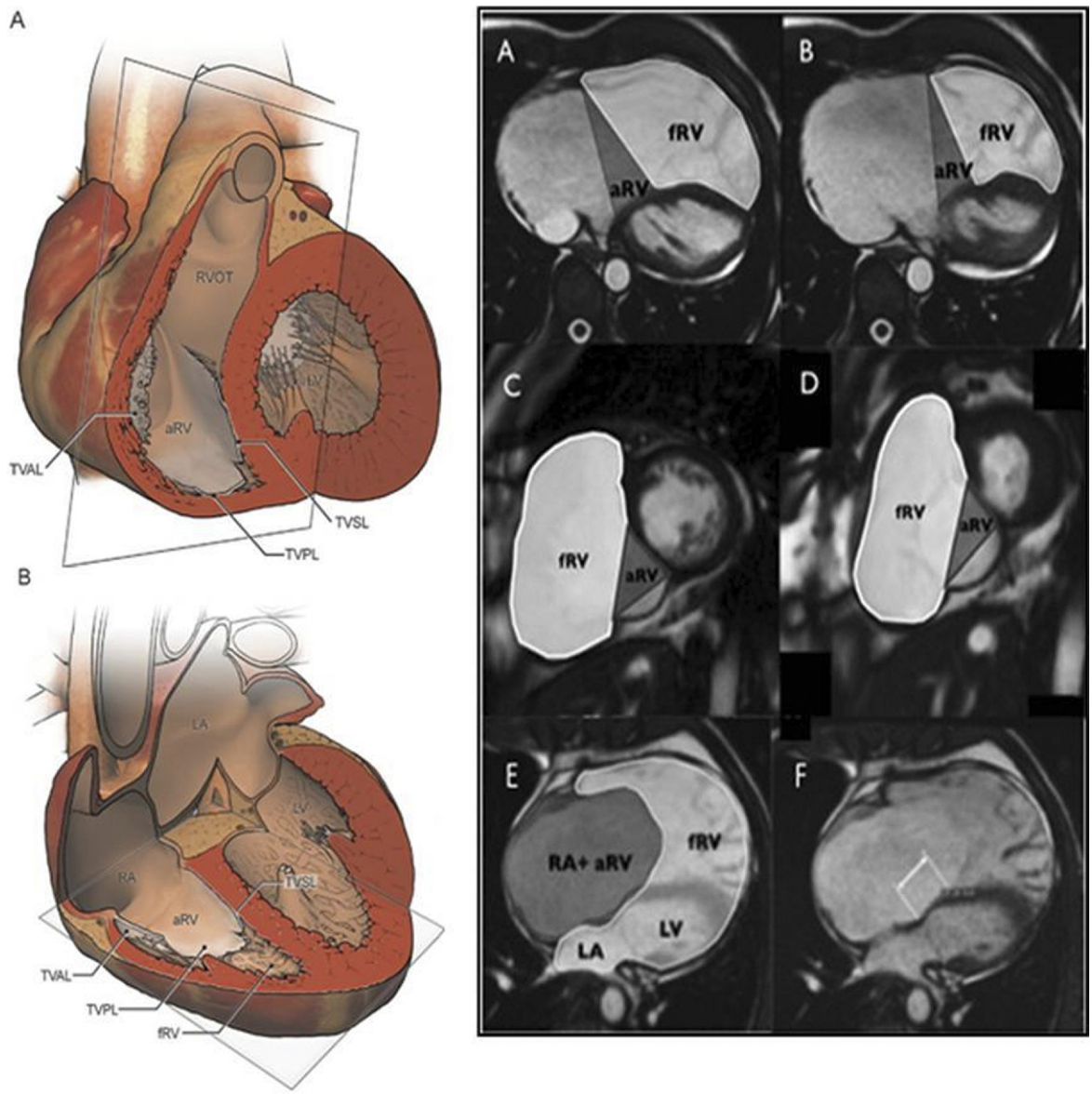


**Figure 3** Left: Three-dimensional reproduction of heart with Ebstein malformation demonstrating: views of tricuspid valve from (A) short axis and (B) axial imaging. Right: Cardiac MRI showing systolic and diastolic contours of functional RV and atrialized portion of RV in (A,B) axial and (C,D) short-axis views. (E) Severity index representing ratio of areas of right atrium and atrialized RV in numerator and summation of functional RV and left atrium and left ventricular areas in denominator (ie, severity index = [right atrial area + atrialized right ventricular area]/[functional right ventricular area + left atrial area + left ventricular area]). (F) Degree of apical displacement of septal leaflet of tricuspid valve (in millimeters) measured in ventricular diastole. aRV, atrialized right ventricle. fRV, functional right ventricle; LA, left atrium; LV, left ventricle; RA, right atrium; TVAL, tricuspid valve anterior leaflet; TVPL, tricuspid valve posterior leaflet; TVSL, tricuspid valve septal leaflet. (Reprinted with permission.)

ered as an alternative to two right-sided valve replacements, particularly when the RV is on the smaller side.

### Tricuspid Regurgitation in Association with Right Ventricular Outflow Tract Obstruction (TOF, Pulmonary Stenosis or PA with Ventricular Septal Defect)

The free pulmonary regurgitation that results from relief of right ventricular outflow tract obstruction, as an essential component of the complete surgical repair of TOF, pulmonary stenosis, and PA with ventricular septal defect (VSD), can result in volume overload with secondary right ventricular dilatation and the development of functional TV regurgitation.[13] TR can also result from leaflet distortion associated with VSD closure in such cases. PVR is indicated in this group of patients in the presence of symptoms, RV volume overload, or the development to arrhythmias (atrial or ventricular). Concomitant TV repair at the time of PVR is still controversial because it is unclear the effect of PVR alone on RV size and secondary TR. In the series reported by Kogon et al,[14] the authors analyzed 35 patients who underwent PVR subsequent to repair of TOF or congenital pulmonary ste-

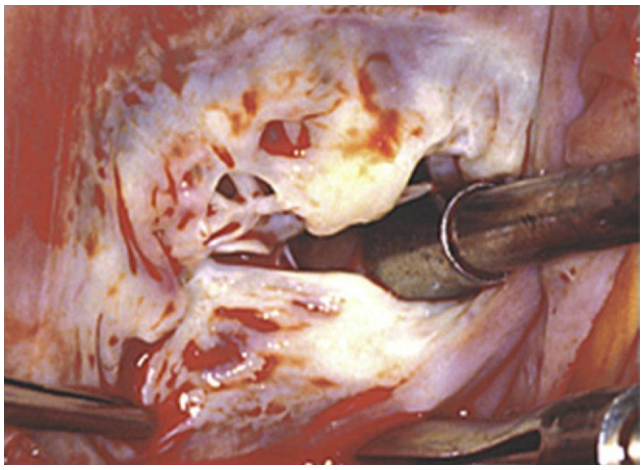

**Figure 4** Intraoperative photo showing dysplastic tricuspid valve in the setting of PA with IVS. (Reprinted with permission from the Mayo Foundation for Medical Education and Research.)

nosis in the period between 2002 and 2008. These patients had at least moderate TR at the same time of their PVR. The degree of TR as well as RV dilatation decreased significantly at 1 month postoperatively, with no further improvement at late follow-up.

We analyzed the outcomes of 53 patients who had surgical treatment of pulmonary valve stenosis between 1951 and 1982.[15] In 28 patients, there were 35 re-interventions, which included PVR for free pulmonary regurgitation in 21 patients, open valvotomy in five, and pulmonary balloon valvuloplasty in three for residual pulmonary valve stenosis, closure of atrial septal defect in two, right ventricular outflow tract reconstruction in one, closure of iatrogenic VSD in one, ligation of aortopulmonary fistula in one, and tricuspid valve annuloplasty with simultaneous coronary artery bypass grafting in one patient. In addition, atrial and ventricular arrhythmias were common, occurring in 20 patients (38%). Severe pulmonary valve regurgitation, although well tolerated initially, has deleterious effects on RV systolic function and exercise capacity over time. In addition, evidence suggests that long-standing pulmonary regurgitation with right-sided cardiac chamber enlargement may be associated with the development of arrhythmias and sudden death.[16] In our practice, we have used the development or progression of TR related to annular dilation as an indication for PVR. In addition, it is our policy to repair the TV when TR is graded moderate or more. Importantly, the decision to repair the TV should be made preoperatively because the conditions of anesthesia generally reduce the grade of TR by at least one grade.

There were a total of 446 reoperations for conduit failure in 370 patients in the period from 1964 to 2001.[17] Progression of TR is an indication for conduit replacement in this group of patients. We have a low threshold for tricuspid valve repair under these circumstances, and it is considered at the time of the first conduit change or concomitant with PVR. TV repair represented 5.7% of the total associated procedures that were performed at the initial time of the conduit operation during the same time period.[17] Progression of TR is considered as a marker of the progression of conduit dysfunction and the need for surgical intervention to prevent the occurrence of arrhythmias and the progression of ventricular dysfunction and right-sided heart failure. The importance of continued follow-up for all patients on a regular basis cannot be overemphasized so that the timing of reoperation is optimized. In general, we prefer a flexible ringed annuloplasty for tricuspid repair when somatic growth is complete and utilize eccentric annuloplasty techniques in earlier childhood.

### Tricuspid Regurgitation in Association with VSD

VSD is one of the most frequently encountered congenital cardiac anomalies. TR in association with VSD can result from various mechanisms which may include: (1) leaflet deformity, (2) TV overriding or straddling, (3) abnormal chordal attachments (Fig. 5), (4) clefts in or between the tricuspid valve leaflets, (5) Gerbode defect, (6) leaflet distortion secondary to patch closure of VSD, and (7) TV detachment. Detachment of the TV septal and/or anterior leaflet is sometimes required to facilitate VSD closure, which in turn may reduce the chance of residual shunt or heart block. The concern with this technique is related to the potential for TV dysfunction and regurgitation, but the data are few and long-term outcomes related to the TV function are unknown. Theoretical impairment of the TV annulus in the long term may also exist, which can result in valvular stenosis.[18] Bol-Raap et al[19] reported a series of 188 patients who underwent VSD closure between 1992 and 2001. Temporary detachment of the TV was performed in 46 patients (24%). Trivial TR was present postoperatively in 10 patients (22%) among the detachment group, and in 57 patients from the non-detachment group (40%). No patient developed tricuspid stenosis. Mean follow-up was 2.6 years, and no patient required reoperation for TV dysfunction. Although controversial, the study reported by Tatebe et al[20] recommended avoidance of detachment of the TV in small infants and those with Down syndrome because of the high incidence of TR. However, we agree with Bol-Raap et al that VSD closure can be performed

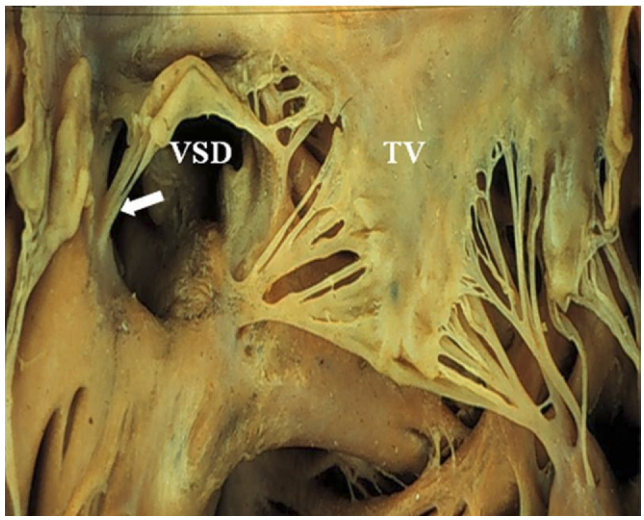


**Figure 5** Pathologic specimen showing abnormal chordal attachment (*arrow*) of the tricuspid valve in relation to the VSD. TV, tricuspid valve; VSD, ventricular septal defect. (Reprinted with permission from the Mayo Foundation for Medical Education and Research.)

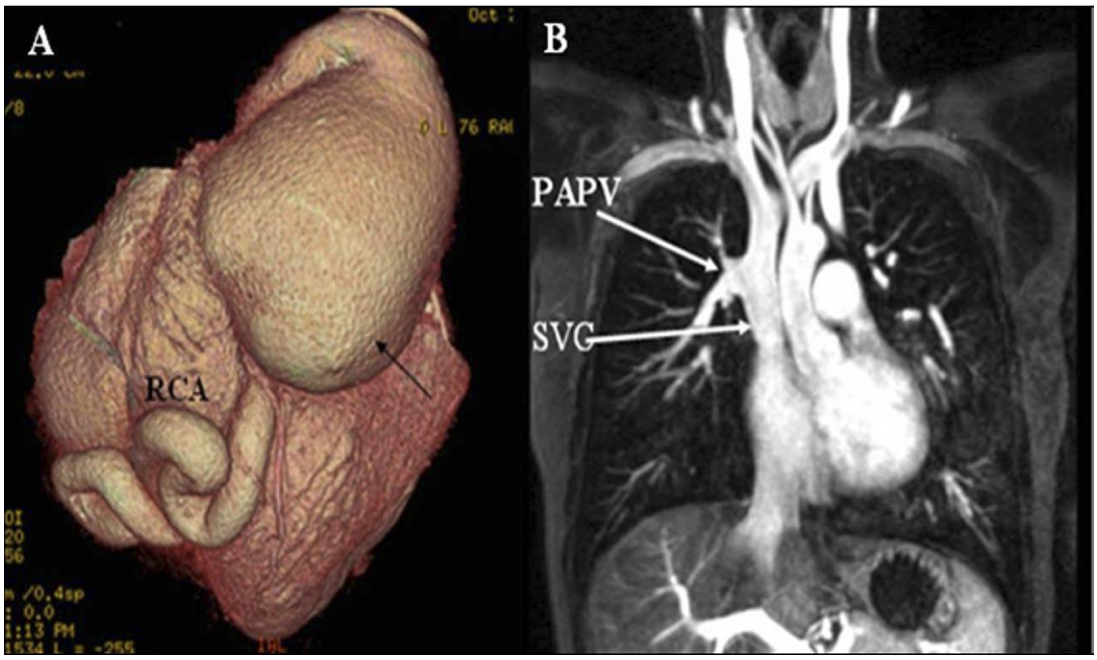


**Figure 6** (A) Cardiac CT scan with 3D reconstruction showing giant right coronary artery aneurysm (*black arrow*) in the setting of congenital right coronary artery-to-coronary sinus fistula. (B) Cardiac MRI showing partial anomalous pulmonary venous connection of the right upper pulmonary vein to the superior vena cava. In A or B, the tricuspid valve annulus is usually dilated secondary to right ventricular enlargement and volume overload. RCA, right coronary artery; PAPV, partial anomalous pulmonary vein; SVCL, superior vena cava. (Reprinted with permission of the Mayo Foundation for Medical Education and Research.)

safely with a very low complication rate, and TV detachment does not result in an increased incidence of TR.

### The Gerbode Defect

Left ventricular-to-right atrial shunts represent less than 1% of congenital heart diseases.[21] In 1958, Franke Gerbode reported the first series of successful repairs of such anomalies.[22] The septal leaflet of the TV divides the membranous septum into interventricular and atrioventricular (AV) portions. The defect can occur in any or both of these portions; the septal leaflet of the TV is commonly involved. If the shunt is in the interventricular segment of the membranous septum, it can result in significant TR. This often requires repair at the time of VSD closure.

### Tricuspid Regurgitation Secondary to Annular Dilatation and Right Ventricular Volume Overload

Congenital coronary arterial fistulae (Fig. 6A), atrial septal defect, and partial anomalous pulmonary venous connection (Fig. 6B) represent examples of congenital lesions that are characterized by large left-to-right shunt. The resultant large RV volume overload leads to RV enlargement with secondary TV annular dilatation. The end result may be the development of significant TR.

### TR Secondary to Implantable Permanent Pacemaker or Cardioverter-Defibrillator Leads

The use of permanent pacemakers (PPM) and implantable cardioverter-defibrillators (ICD) in the congenital population

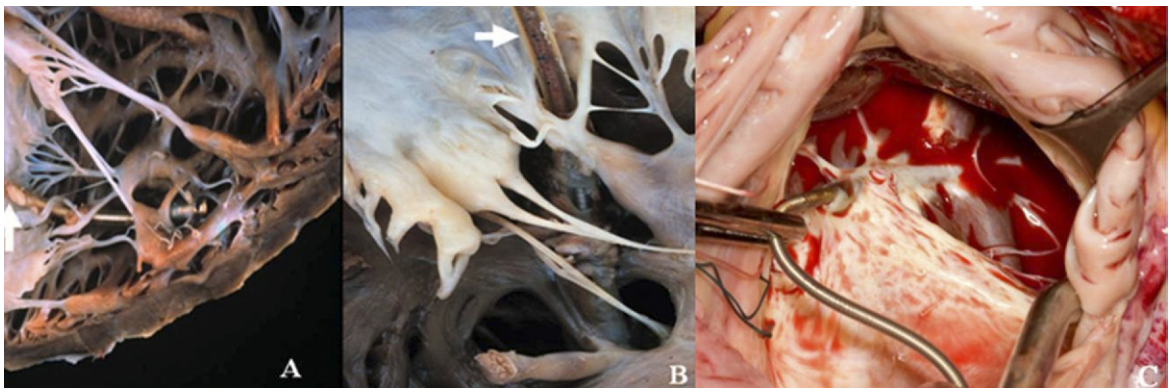


**Figure 7** Pathologic specimens (A-B) and intraoperative photo (C) showing perforation or entanglement of the tricuspid valve leaflet with pacemaker or cardioverter-defibrillators lead (*arrows*). (Reprinted with permission from the Mayo Clinic.)

is increasing because both atrial and/or ventricular arrhythmias are a frequent late complication.

Consequently, TR secondary to these devices are increasing. The mechanisms of TR (Fig. 7A-C) are related to: (1) leaflet perforation by the lead; (2) lead entanglement of the TV apparatus; (3) lead impingement of the TV leaflets; and (4) lead adherence to the TV. In our experience, we analyzed the records of 571 consecutive patients with non-Ebstein TR who underwent TV operation for severe TR in the period between 1993 and 2010. Twenty-five patients (4%) had severe TR secondary to PPM or ICD lead placement. TV replacement was performed in 14 patients (56%), while it was repaired in the remaining 11 patients (44%).

## Indications for Surgery

TR has been considered a benign lesion for a long time, but recent studies suggest that irrespective of pulmonary artery pressure or left ventricular ejection fraction, TR negatively affects long-term survival.[23] The reported 2-year postoperative event-free survival in the presence of severe preoperative RV dysfunction is 57%.[24] Severe TR induces chronic RV volume overload, which leads to progressive ventricular dilatation, dysfunction, and eventually right-sided heart failure. Timely correction of TR will preserve RV function, improve functional capacity, and improve long-term survival. TV repair is the preferred treatment strategy when it is feasible, particularly in children. Optimal timing is now recommended before the onset of RV dysfunction, even in asymptomatic patients.[25,26]

The indications for surgery (summarized in Table 2) in TR include symptoms or cyanosis (when atrial septal defect is present), decreased exercise tolerance, progressive cardiomegaly on chest x-ray, progressive RV dilatation or reduction of RV systolic function by echocardiography, or appearance of atrial or ventricular arrhythmias. In borderline situations, the echocardiographic determination of high probability of TV repair makes the decision to proceed earlier with operation easier.

Progression of TR is taken into consideration for timing of intervention for PVR as well (Table 2). In general, we repair the TV when the degree of TR is moderate or more at the time of PVR. It is important to make the decision for TV intervention on preoperative outpatient echocardiography rather than on intraoperative transesophageal echocardiography because of the difference in hemodynamics under general anesthesia.

**Table 2 Indications for Surgery**

| |
|---|
| **A. Tricuspid Regurgitation** |
| Symptoms or reduced exercise intolerance |
| Cyanosis (concomitant interatrial communication) |
| Progressive RV dilatation |
| Progression of RV dysfunction |
| Onset, progression of arrhythmias |
| **B. Pulmonary Valve Replacement** |
| Symptoms of right heart failure and exercise intolerance |
| RV systolic pressure > 2/3 of systemic blood pressure |
| RVEDVI ≥ 150 ml/m$^2$ |
| Onset, progression of arrhythmias |
| Prolongation of QRS complex ≥ 180 msec or > 3.5 msec/year |
| Progressive RV dysfunction |
| Progression of TR |

## Surgical Techniques

### Tricuspid Valve Repair

The goal of the operation is repair of the TV. To increase the number of successful TV repairs, particularly in children, we have been using a cone-type reconstruction (Fig. 8A-J).[27,28] This results in 360° of tricuspid leaflet tissue surrounding the right AV junction. This allows leaflet tissue to coapt with leaflet tissue, similar to what occurs with normal TV anatomy. The hinge point of the reconstructed TV is at the true TV annulus (AV junction). Modifications include suturing the base of the intact papillary muscle on the RV free wall to the ventricular septum (Sebening stitch) at the appropriate level with interrupted, pledgeted mattress sutures (Fig. 9).[29] It is important to place these mattress sutures deep into the ventricular septum and that the anterior leaflet has been thoroughly mobilized (delaminated). Alternatively, a monocusp-type repair based on a large anterior leaflet has also been successful.[30,31] It is important that the repair is tension free. To facilitate this, augmentation of the mid-anterior leaflet with an ellipse of autologous pericardium or other pliable synthetic material as CorMatrix (Fig. 10) can increase the height of the anterior leaflet, which facilitates coaptation with the ventricular septum while minimizing tension at an inferior annuloplasty line. Importantly, we avoid the creation of complete pericardial leaflets (from annulus to leading edge) because of poor durability. When inferior leaflet tissue is absent, this area is often the site of residual regurgitation, but this area can be bridged with a piece of autologous pericardium (Fig. 11A-B). If no septal leaflet is found, we perform the surgical delamination of the anterior and inferior leaflets as usual, and then the papillary muscle(s) is mobilized and sutured to septum. The anterior leaflet is rotated clockwise toward the septum, and the inferior leaflet is sutured to the septum beyond the coronary sinus instead of the septal leaflet. In case of complete tethering of the leading edge of the leaflet (ie, linear attachment; Fig. 12A-B), surgical fenestrations are performed distally, which effectively result in the development of chordae (Fig. 12C). Annular plication sutures, purse-string annuloplasty, or eccentric commissuroplasty can be performed to further narrow the tricuspid annulus. A *flexible* annuloplasty C-shaped ring is our preference and is used routinely when somatic growth allows. This begins at the anteroseptal commissure and ends at the inferoseptal commissure (adjacent to the coronary sinus).

The addition of an edge-to-edge stitch, thus creating a double orifice TV, can be added to the above maneuvers to optimize coaptation and improve valve competency in selected circumstances provided iatrogenic stenosis is avoided. Closure

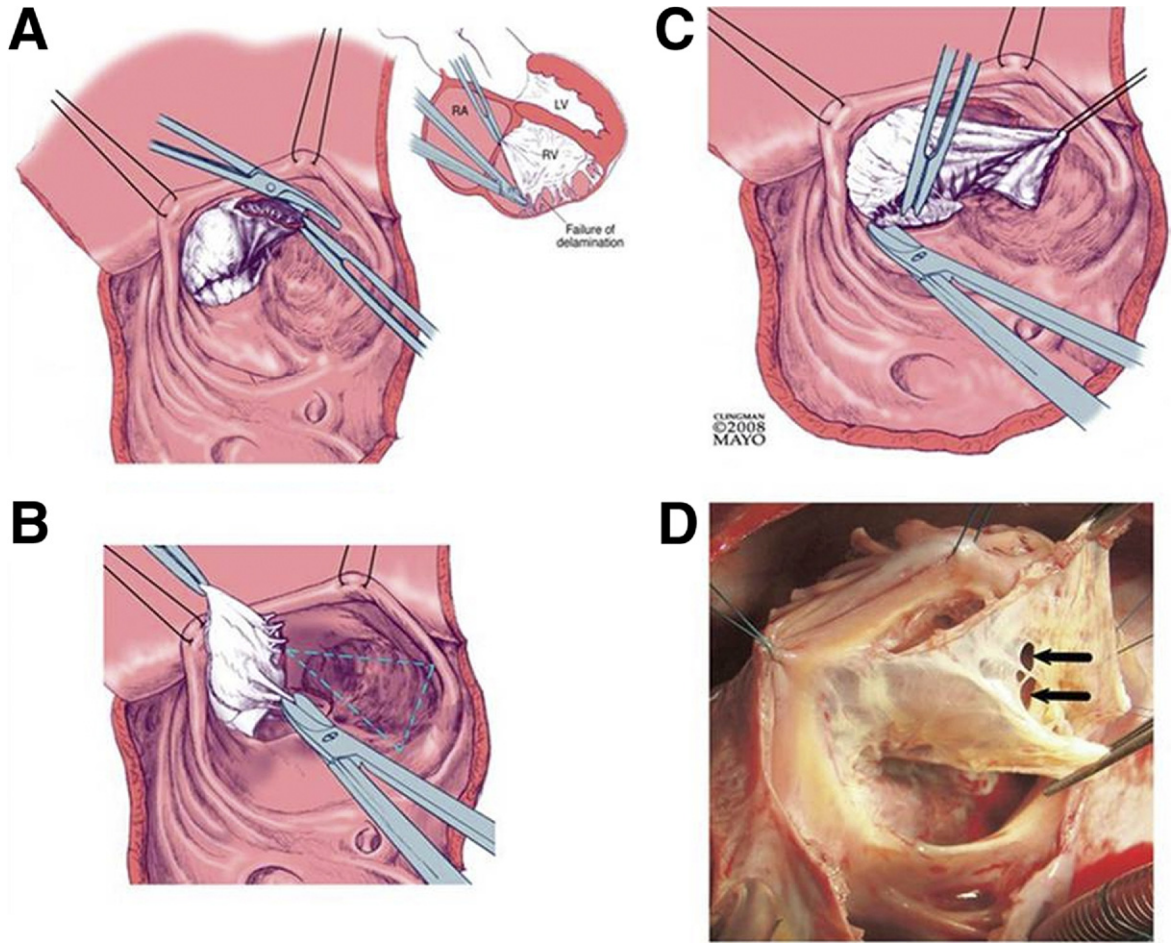


**Figure 8** Operative steps for the "cone technique." (A) The first incision is made with a no. 15 blade in the anterior leaflet at 12:00; the incision is a few millimeters away from the true annulus. The incision is then extended rightward in a clockwise fashion using a scissors. It is common for there to be a true space between the anterior leaflet and the RV in this region (ie, normally delaminated leaflet). However, when the transition is met between the anterior and inferior (posterior) leaflets, it is common for there to be failure of delamination (*inset*), resulting in fibrous and muscular attachments between the leaflet and myocardium. The diagram demonstrates the scissors approaching the area where there is some adherence of leaflet tissue to the underlying myocardium. The dissection continues in a way that a portion of distal anterior leaflet and some inferior leaflet tissue is "surgically delaminated." The most important aspect of this surgical delamination is to incise all fibrous and muscular attachments between the body of the leaflet and the right ventricular myocardium, but to maintain intact all fibrous and (and occasionally muscular) attachments of the leading edge of the leaflet to the underlying myocardium. Importantly, do not disrupt chordal attachments to the leading edge of any leaflet. (B) As the anterior and surgically delaminated inferior leaflet is reflected away from the right ventricular myocardium, all fibrous and muscular attachments into the body of the underside of the leaflet are incised as shown with the scissors. It is important to keep all attachments of the leading edge of the leaflet intact; if the edge is linearly attached, then surgical fenestrations are created distally. The dotted triangle represents the atrialized RV. (C) Dissection is continued with a scissors, with the goal of taking down all attachments between the septal leaflet and myocardium but preserving all attachments of the leading edge to the endocardium, as described above. The dissection should proceed medially all the way to the anteroseptal commissure. The leaflet tissue is typically very fragile and thin in this area. There can be marked variability in the status of the leading edge of the septal leaflet as was described for the anterior and inferior leaflets. If there is a linear attachment, then surgically created fenestrations are also made in this leaflet (not shown). (D) Intraoperative photo demonstrating the mobilized anterior and inferior (posterior) leaflets. Natural fenestrations are shown at the junction of the anterior and inferior leaflets (*arrows*). LV, left ventricle; RA, right atrium; RV, right ventricle. (*Figure continues.*)

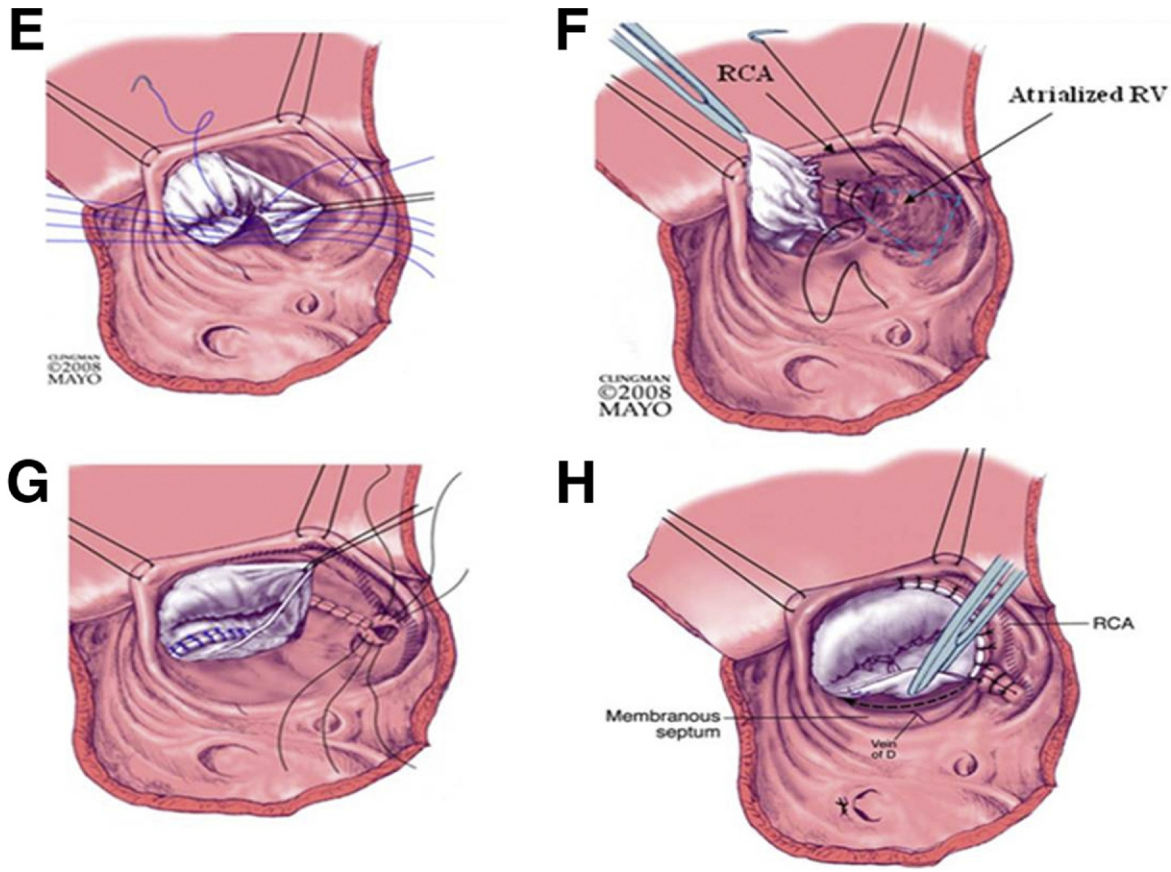


**Figure 8** (E) After the anterior, inferior, and septal leaflets have been completely mobilized, the cut edge of the inferior leaflet is rotated clockwise to meet the proximal edge that has been prepared of the septal leaflet. The two are approximated with interrupted 6-0 monofilament sutures completing the cone reconstruction. This results in 360° of leaflet tissue that will make up the new tricuspid valve orifice. (F) After the cone reconstruction is completed, the atrialized RV (RV) is examined to determine if plication is necessary. Note the position of the right coronary artery (RCA) in the true tricuspid valve annulus and keep in mind that there are acute marginal branches of the right coronary artery that can also be compromised with plication. This figure demonstrates the technique for internal plication of the atrialized RV. Monofilament 4-0 or 5-0 is utilized and the suture is begun distally, ie, closest to the apex of the RV. It is important to frequently examine the outside of the inferior wall of the RV to insure that inadvertent compromise of branches of the right coronary artery is avoided. (G) The suture line is advanced toward the base of the heart, ie, toward the AV groove. As the dotted lines of the triangle are effectively approximated, the atrialized RV is excluded. Occasionally, the suture line extends beyond the AV groove onto the right atrium. Care is taken to avoid compromise of the right coronary artery. After the sides of the triangle are approximated, the entrance into the excluded atrialized segment of the RV is then closed to eliminate the "blind pouch." (H) After the plication or resection is completed, the newly constructed tricuspid valve is then reattached below the level of the true tricuspid annulus (ventricular side of conduction tissue). Because the neotricuspid valve will have an orifice that is smaller than the original dilated AV junction, a plication of the inferior annulus is necessary to meet the size of the neotricuspid valve. The inferior annulus is usually plicated with two to four simple or figure-of-eight 5-0 monofilament sutures. Proper placement of these sutures is critical to the success of this repair. The sutures need to be deep enough to maintain the integrity of the new annulus intact *without* compromising or significantly distorting the right coronary artery. If the size discrepancy between the true tricuspid annulus and the neotricuspid valve is large, smaller annular plication sutures should be placed in multiple areas around the true annulus to avoid distortion of the right coronary artery by a large plication in a single area. To avoid heart block, the suture line is deviated caudad to the membranous septum and AV node, which is marked by the previously mentioned vein and fatty tissue. *(Figure continues.)*

of intra-atrial shunts is performed routinely when operation is performed beyond infancy. Right reduction atrioplasty is routinely performed, and the maze procedure is added when atrial tachyarrhythmias are present.

In cases of TR secondary to VSD closure, there is often disrupted commissural chordae (Fig. 13A) at the anteroseptal commissure with or without annular dilatation. Treatment consists of approximating the adjacent edges of the leaflets at this commissure together, thus obliterating that commissure. In the presence of annular dilatation, a flexible annuloplasty ring can be added to remodel the annulus as described above (Fig. 13B).

In the presence of AV septal defect (Fig. 14A-B), closure of the residual cleft in the left AV valve is performed, and placement of annuloplasty rings on each valve is routine. In selected situations, leaflet augmentation with pericardium or

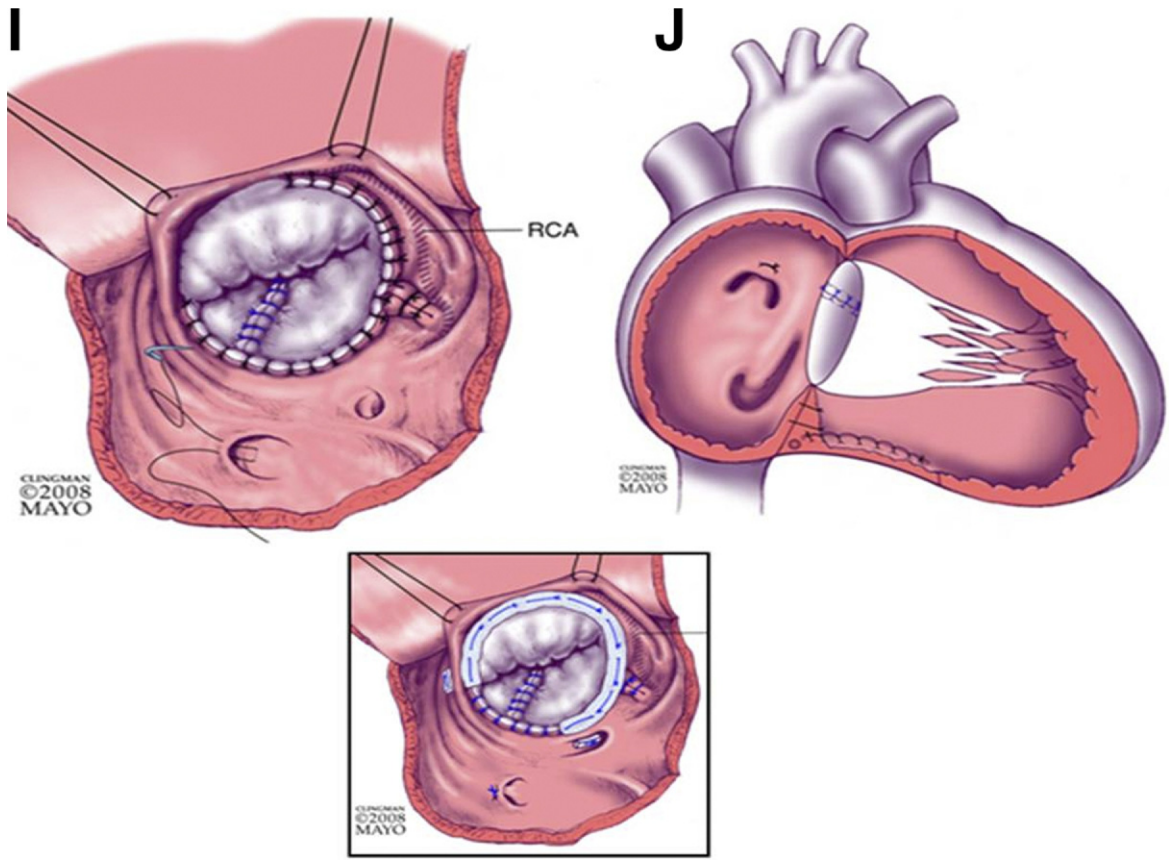


**Figure 8** (I) The completed cone reconstruction of the tricuspid valve. Saline is injected via bulb syringe into the RV to examine competency of the tricuspid valve. Any residual fenestrations or areas of leak are repaired as needed. A subtotal closure of the patent foramen ovale or atrial septal defect is usually performed. If it is believed that a bidirectional Glenn shunt is needed because of a small effective orifice of the neotricuspid valve, or because of severely depressed RV function, then the intra-atrial communication may be closed completely. Redundant right atrium is excised from each side of the atriotomy and then the atriotomy is closed. RCA, right coronary artery. (*Inset*; ring annuloplasty to reinforce the repair). (J) The completed cone reconstruction of the tricuspid valve for Ebstein's anomaly. This represents an "anatomic repair" because there is 360° of tricuspid leaflet tissue that surrounds the orifice of the tricuspid valve that is anchored at the level of the normal right AV junction (true tricuspid valve annulus). Extreme forms of thinned, atrialized RV are plicated and redundant right atrium is excised. (Reprinted with permission of Mayo Foundation for Medical Education and Research.)

other prosthetic material is used to increase the height of the attenuated leaflet.

In the presence of TR induced by PPM or automatic implantable cardioverter defibrillator (AICD) leads, repair techniques vary and depend on the degree of damage of leaflet(s). In the absence of extensive TV leaflet damage, valve repair is preferred and usually involves: (1) removing (incising) the lead away from the damaged leaflet; (2) suture repair of the leaflet defect if present; or (3) repositioning the lead by suture fixation in the recess of either the infero-septal or antero-inferior commissure (Fig. 15A-C). Ringed annuloplasty is performed. We confirm the resolution or improvement of the TR with intraoperative transesophageal echocardiography.

### Tricuspid Valve Replacement

Bioprosthetic TV replacement remains a good alternative for the treatment of TR when valve repair is not feasible, particularly in adults. Porcine bioprosthetic valve replacement is preferred because of relative good durability of the porcine bioprosthesis in the tricuspid position and the lack of need for chronic warfarin anticoagulation. Pericardial bioprostheses should be avoided in the tricuspid position because of relatively stiffer leaflets and poor durability in this position. Mechanical TV replacement should be considered rarely and only in selected circumstances because there is a higher frequency of prosthetic valve dysfunction (thrombosis) compared with mechanical valves in other cardiac positions, particularly when right ventricular function is poor and disc mobility may be reduced.

When TV replacement is required, care is taken to preserve the conduction tissue and avoid injury of the right coronary artery. The conduction tissue is usually identifiable by a small vein traversing an area of yellow (fat) tissue adjacent to the membranous septum. Before replacing the TV, we resect the portion of the anterior leaflet toward the RV outflow tract to avoid potential RV outflow tract obstruction after TV replace-

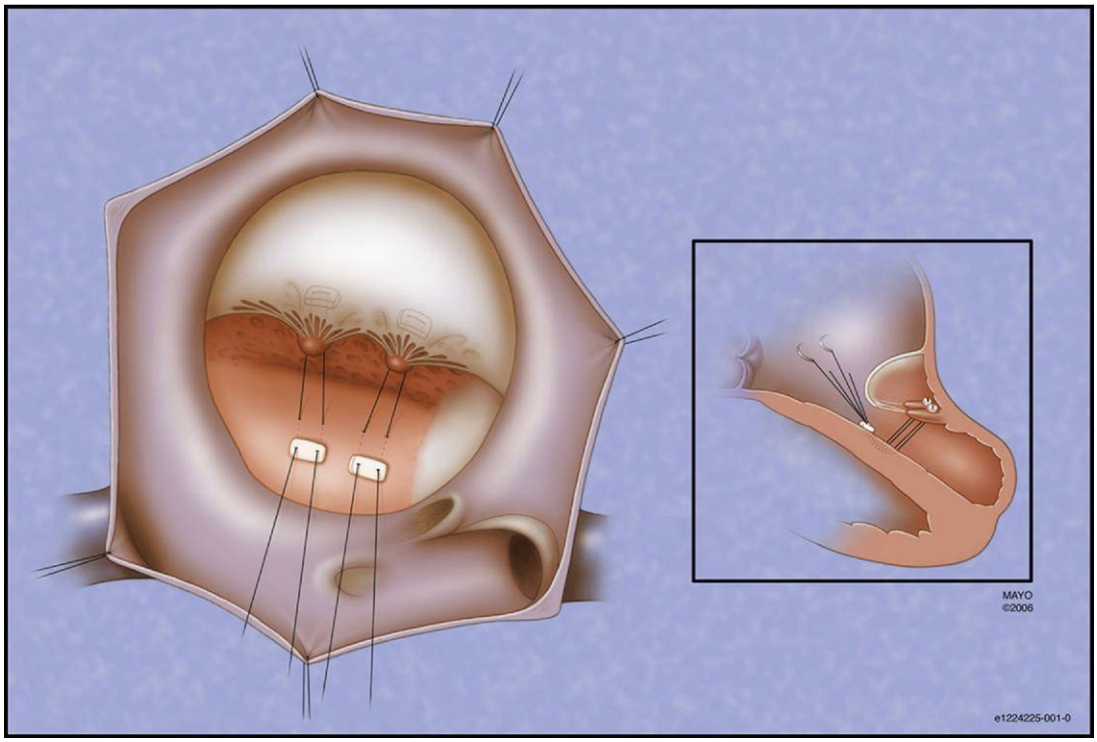


**Figure 9** Diagram showing the Sebening stitch in which the base of the intact papillary muscle on the right ventricular free wall is moved toward the ventricular septum at the appropriate level with interrupted, pledgeted mattress sutures. It is important to place these mattress sutures deep into the ventricular septum. In addition, it is important to insure thorough mobilization of the anterior leaflet to avoid dimpling of the right ventricular free wall (*inset*). (Reprinted with permission from the Mayo Clinic.)

ment. In EM, the suture line is deviated toward the atrial side of the true annulus and membranous septum to avoid injury to the AV node. Posteriorly, the suture line is typically in the atrial septum (often incorporating the edge of the atrial septal defect patch) (Fig. 16A-B). To avoid injury to the right coronary artery (Fig. 17), the anterior suture line is deviated cephalad to the true annulus, where the smooth and trabeculated portions of the atrium meet each other. The bioprosthesis is oriented so the recess between the struts straddles, the membranous septum, and conduction tissue. We prefer tying the valve sutures while the heart is beating to detect any rhythm abnormalities. The position of the coronary sinus relative to the prosthesis depends on the distance between it and the AV node. The coronary sinus is left draining into the right atrium when there is sufficient distance between it and the conduction tissue. When the coronary sinus is in close proximity to the conduction tissue, the prosthesis is positioned cephalad to it so that the coronary sinus drains below the prosthesis into the RV.

In non-Ebstein congenital TR, all sutures are placed in native anterior and inferior TV leaflet tissue circumferentially. The posterior suture line is typically placed through septal leaflet tissue at the level of the AV groove when sufficient septal leaflet is present; if there is a lack of septal leaflet, suture placement is as described above for EM. Care is taken for accurate suture placement at the anteroseptal commissure, a common site for periprosthetic leak or injury to the conduction system resulting in heart block.

In cases of TV leaflet damage secondary to PPM/AICD leads, tricuspid valve replacement is performed as described above for non-Ebstein congenital TR. The decision to remove the endocardial lead and place an epicardial system is individualized. In circumstances when there are excellent thresh-

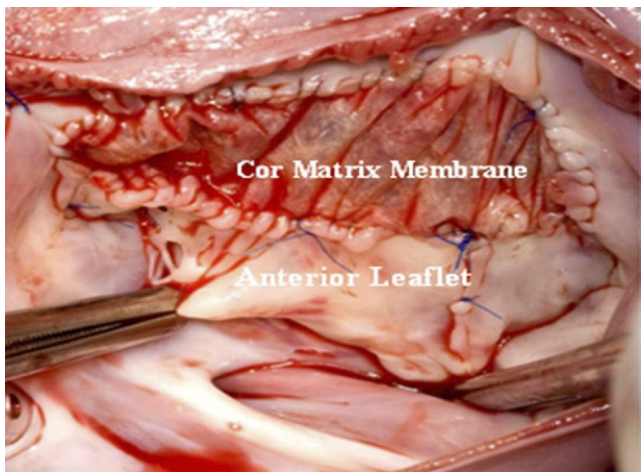


**Figure 10** Intraoperative photo showing the augmented anterior tricuspid valve leaflet with cor matrix membrane, which increases the height of the anterior leaflet, and facilitates coaptation with the ventricular septum and or septal leaflet while minimizing tension at an inferior annuloplasty line. Importantly, we avoid the creation of complete pericardial leaflets (from annulus to leading edge) because of poor durability.

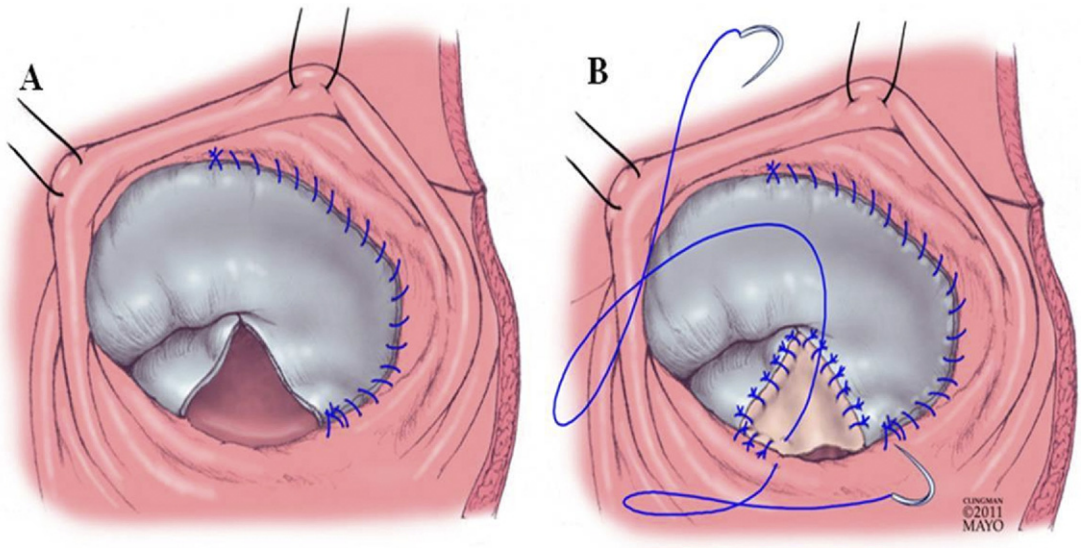


**Figure 11** (A-B) When inferior leaflet tissue is absent or septal leaflet is diminutive, this area can be bridged with a piece of autologous pericardium to avoid iatrogenic tricuspid stenosis. Interrupted sutures are often used to avoid a "purse string effect." (© 2011 Mayo Clinic.)

olds in the endocardial system and there is significant tricuspid annular dilatation, the ventricular lead can be maintained intact and positioned lateral to the sewing ring of the bioprosthesis. It is important to be sure the lead is freely mobile alongside the prosthesis.

In general, postoperative management includes short-term (3 months) warfarin anticoagulation for porcine bioprostheses and life-long aspirin, 81 mg daily. When a mechanical valve is used, the target INR is 3 to 3.5 in addition to aspirin, 81 mg daily.

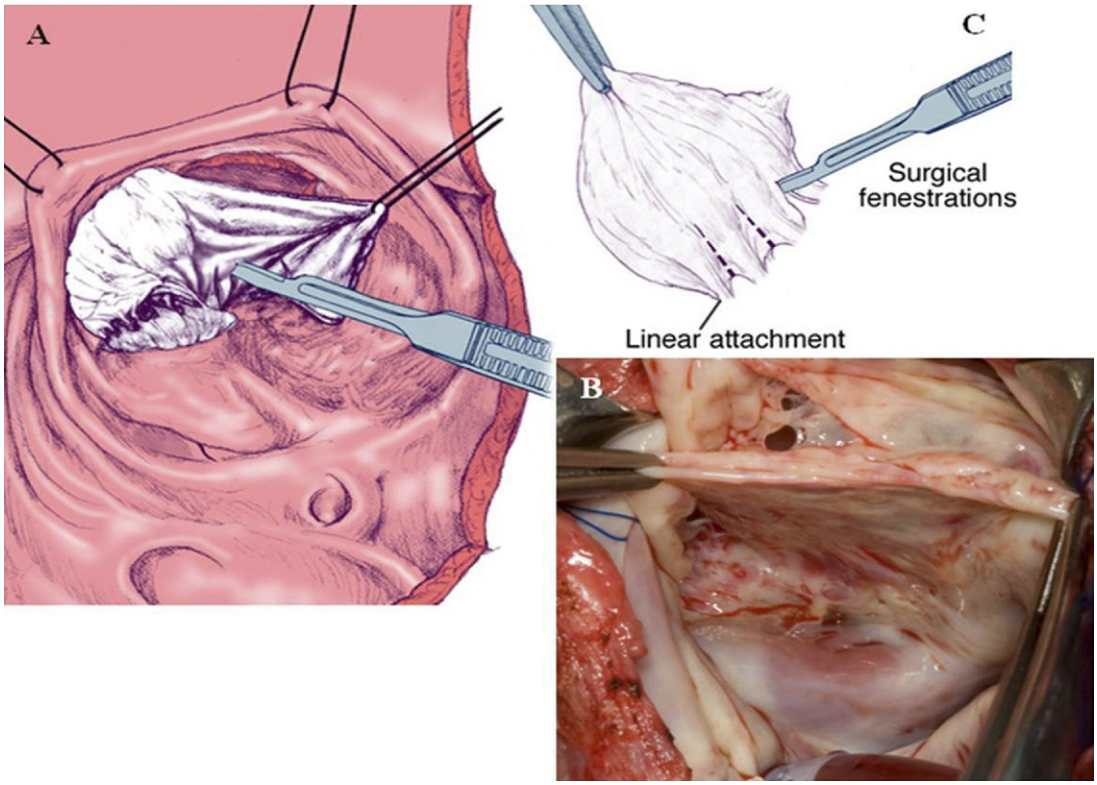


**Figure 12** In case of complete tethering of the leading edge of the leaflet, ie, linear attachment (A,B), surgical fenestrations are performed distally that effectively result in the development of chordae (C). (Reprinted with permission from the Mayo Clinic.)

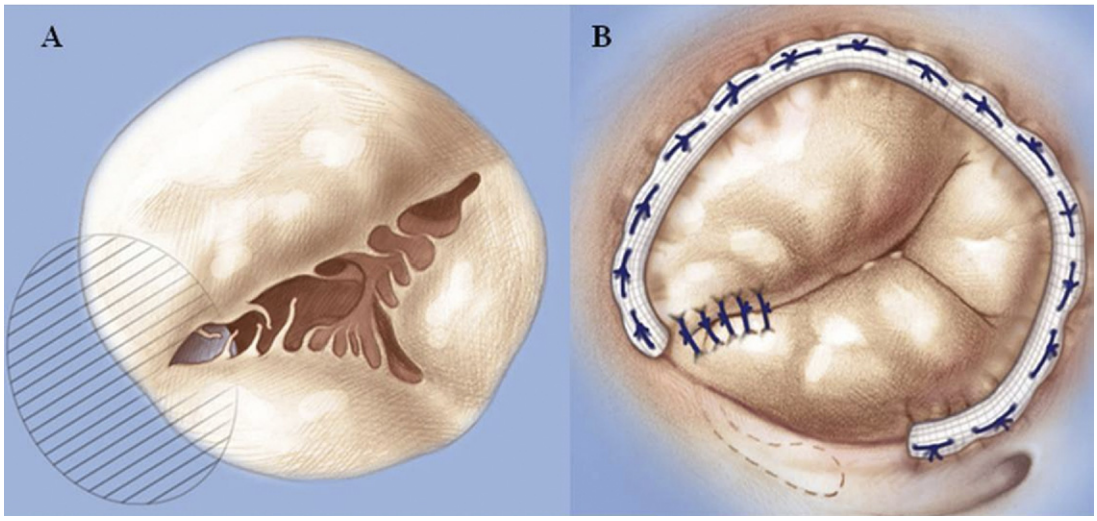


**Figure 13** In cases of tricuspid regurgitation secondary to VSD closure, there is often disrupted commissural chordae (A) at the anteroseptal commissure with or without annular dilatation. Treatment consists of approximating the adjacent edges of the leaflets at this commissure together, thus obliterating that commissure. In the presence of annular dilatation, a flexible annuloplasty ring should be added to remodel the annulus as described above (B). (Reprinted with permission from the Mayo Clinic.)

## Additional Perioperative Strategies

The following operative and postoperative strategies should be considered following correction of TR, particularly when there is significant RV dysfunction: (1) measures that decrease RV afterload and pulmonary artery pressure, eg, nitric oxide, intra-aortic balloon counter pulsation (especially in adult patients), manipulation of mechanical ventilation with the $PCO_2$ goal of 30 to 35 mmHg; (2) cautious volume administration with a CVP goal of $<$12 mmHg; (3) temporary atrial pacing to achieve faster heart rate of 100 to 120; (4) minimize transfusions; and, finally, (5) delayed chest closure. Inotropic support while separating from bypass usually in-

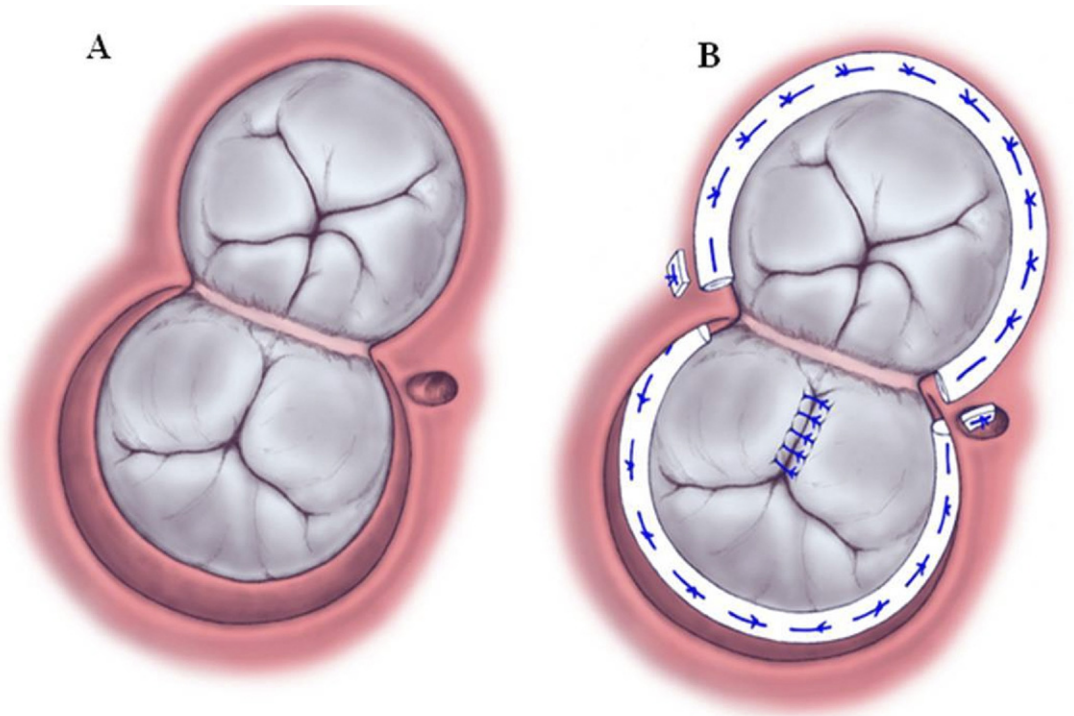


**Figure 14** In the presence of atrioVSD (A), closure of the residual cleft in the left AV valve is performed, and placement of annuloplasty rings on each valve is routine (B). In selected situations, leaflet augmentation with pericardium or other prosthetic material is used to increase the height of the attenuated leaflet (not shown). (Reprinted with permission from the Mayo Clinic.)

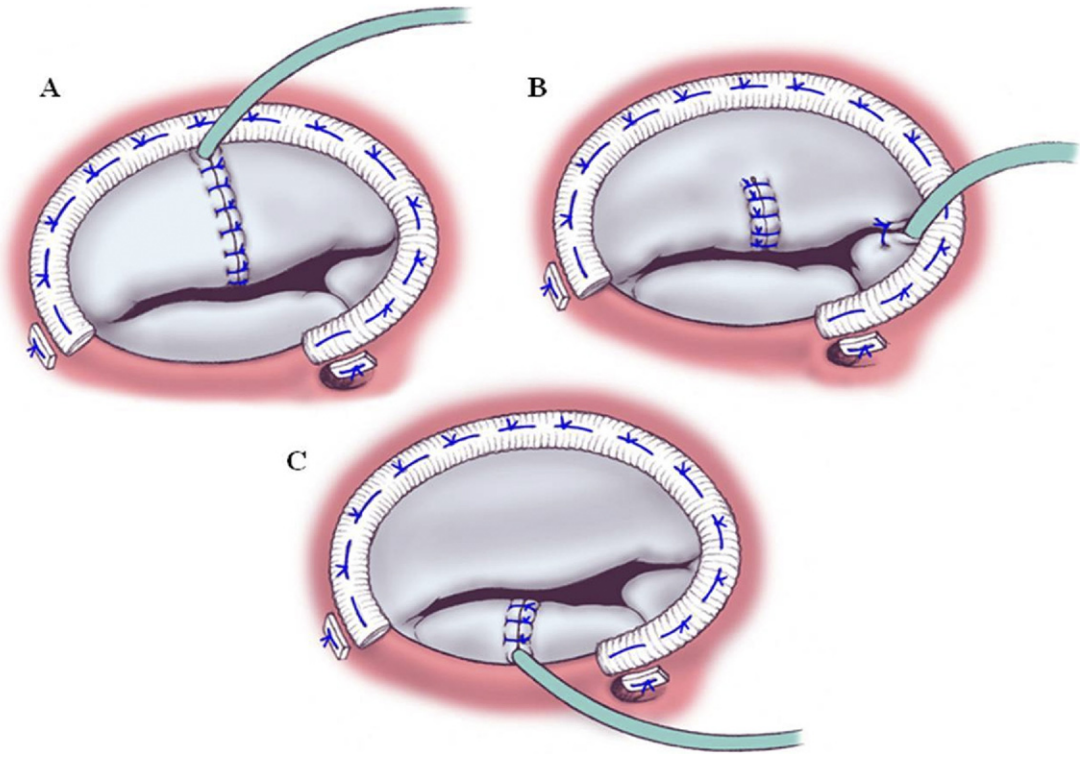


**Figure 15** (A-C) In the presence of pacemaker or cardioverter defibrillator leads, repair techniques vary and depend on the degree of damage of leaflet(s). In the absence of extensive leaflet damage, valve repair is preferred and usually involves: removing (incising) the lead away from the damaged leaflet; suture repair of the leaflet defect if present; or repositioning the lead by suture fixation in the recess of either the infero-septal or antero-inferior commissure, and ringed annuloplasty is performed. (Reprinted with permission from the Mayo Clinic.)

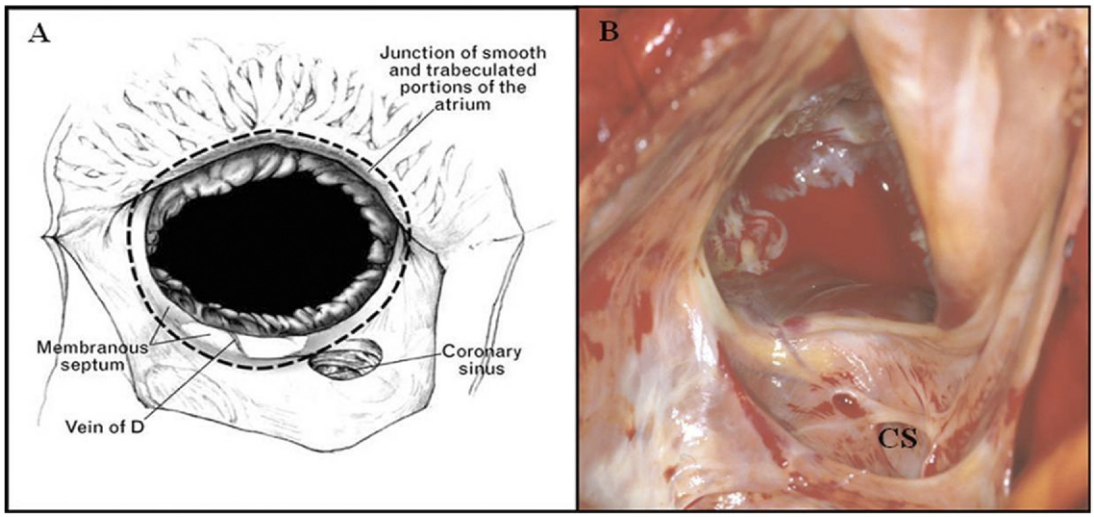


**Figure 16** Diagram (A) with the corresponding intraoperative photo (B) showing the valve suture line (*dotted line*) is placed on the atrial side of the membranous septum and AV node to avoid injury to the conduction system. The suture line is also deviated cephalad to the tricuspid annulus posterolaterally when the tissues are thin, to avoid injury to the right coronary artery. Anteriorly, the suture line is deviated cephalad to the junction of the smooth and trabeculated portions of the right atrium. This results in the prosthesis being mounted in an intra-arterial position. When there is sufficient distance between the coronary sinus and the AV node (marked by the vein of D), the coronary sinus is left draining normally into the right atrium. If the coronary sinus and the conduction tissue are in close proximity, the suture line is deviated further into the right atrium, leaving the coronary sinus to drain below the prosthesis into the RV (not shown). The sutures are placed and tied with the heart perfused and beating to ensure that normal AV conduction is preserved. CS, coronary sinus. (Reprinted with permission from the Mayo Clinic.)

cludes a combination of epinephrine and milrinone infusions. Low doses of vasopressin infusion are often helpful because systemic vasoplegia and low peripheral vascular resistance are commonly present with right-sided heart failure related to TR and/or pulmonary regurgitation. The mechanism of this is not completely understood but may relate to preoperative afterload-reducing agents such as ace inhibitors, or may be related to hepatic congestion, or hormonal abnormalities related to atrial dilatation.

The rate of weaning inotropic support in the postoperative period is related to the degree of RV dysfunction. Inotropic support can be weaned relatively quickly when RV dysfunction is mild to moderate and early extubation is the rule. When there are significant degrees of RV dysfunction, the inotropic wean should be slow because of the expected drop in the cardiac output that can occur between 6 to 12 hours postoperatively.

Although there are no data that afterload reducing agents are helpful for RV dysfunction (in contrast to left ventricular dysfunction), it is common to use them and usually includes ace inhibitors. In addition, beta blockers are also commonly added because of evidence of improvement with left-sided dysfunction. The duration of medical therapy is determined by the primary Cardiologist and is guided by serial echocardiography, exercise testing, etc. In selected circumstance, sildenafil is used for 1 to 3 months when there is known elevation in pulmonary artery pressures with or without severe RV dysfunction. Antiarrhythmic therapy is used as indicated by rhythm abnormalities.

## The Mayo Clinic Experience

From 1993 to 2010, we performed 1,204 procedures for congenital TR. Congenital non-Ebstein tricuspid valve was the etiology in 571 patients. TV repair was the main procedure in 458 patients, at a mean age of 29, while in tricuspid valve replacement was performed in 113 patients (mean age of 40). Table 3 summarizes the different diagnoses for the 571 patients. Repeat sternotomy (≤ 3) was required in 81% of the TV repair group versus 65% in the TV replacement group, while four sternotomies or more were performed in 8% of the TV repair group versus 34% of the TV replacement group. Early mortality in the repair group was 2.4%, while in the replacement group it was 9.7%.

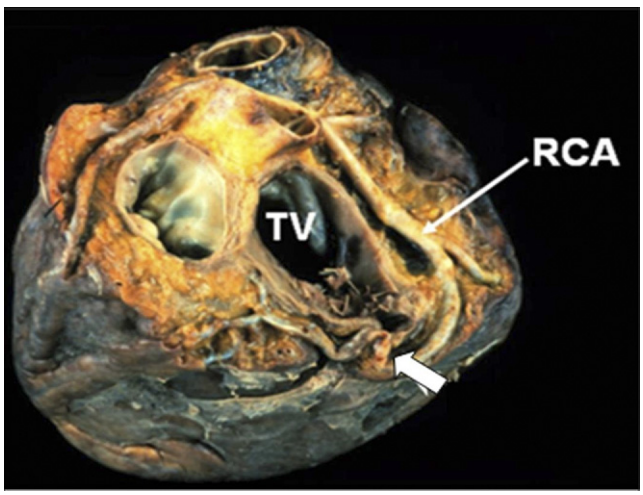


**Figure 17** Pathologic specimen showing the close relation between the right coronary artery and the tricuspid valve annulus. The right coronary artery (*long arrow*) can be easily kinked (*short arrow*) during tricuspid valve repair procedure. RCA, right coronary artery; TV, tricuspid valve. (Reprinted with permission from the Mayo Clinic.)

**Table 3 Etiology of TR in 571 Patients with Non-Ebstein Tricuspid Valve**

| Diagnosis | Percentage |
|---|---|
| TOF/PA with VSD | 31 |
| VSD | 15 |
| AVSD | 14 |
| Transposition of great arteries | 8 |
| Truncus arteriosus | |
| Double outlet right ventricle | |
| Permanent pacemaker | 4 |
| Cardioverter-defibrillator | |
| Congenital, other | 28 |

## Summary

The wide and infinite variability of anatomic abnormality with EM and congenital TV dysplasia demonstrate that every valve is a little different and no two hearts are alike. . .illustrating why this lesion continues to be one of the most challenging valve lesions for the surgeon. There have been more reports in the literature of tricuspid valvuloplasty techniques for EM and congenital tricuspid dysplasia than any other valve lesion in cardiac surgery. While the ability to obtain a competent, durable tricuspid repair has improved in recent years, the surgical treatment of the congenitally abnormal tricuspid valve is still considered palliative because many patients require more than one operation in their lifetime.

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