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# Surgical treatment of Ebstein's malformation: state of the art in 2006

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EBSTEIN'S MALFORMATION IS A CONGENITAL anomaly of the tricuspid valve and right ventricle that is characterized by several features, each of which can exhibit a spectrum of malformation. The first is adherence of the leaflets of the tricuspid valve to the underlying myocardium, this representing failure of delamination during development. This feature involves the septal and inferior leaflets, but rarely the anterior leaflet (Fig. 1). The second feature is anterior and apical rotational displacement of the functional annulus (Fig. 2). The third abnormality is dilation of the "atrialized" portion of the right ventricle, with variable degrees of hypertrophy and thinning of the wall. The fourth finding is redundancy, fenestrations, and tethering of the anterior leaflet. A fifth abnormality is dilation of the right atrioventricular junction, this being the true tricuspid valvar annulus. The final feature is variable ventricular myocardial dysfunction. Each heart with Ebstein's malformation is different, and there is an infinite variability that can occur with the above mentioned characteristics. These anatomical and functional abnormalities cause important tricuspid regurgitation, which results in right atrial and right ventricular dilation, and atrial and ventricular arrhythmias.

## Implications of the disordered anatomy

With increasing degrees of anatomic severity of malformation, the fibrous transformation of leaflets from their muscular precursors remains increasingly incomplete, with the septal leaflet being most severely involved, the inferior leaflet less severely involved, and the anterior leaflet rarely being involved. It is this failure of delamination which moves the hinge points of the septal and inferior leaflets, the functional annulus, into the ventricular cavity, and away from the anatomic right atrioventricular junction, resulting in a rotational or spiral displacement of the functional valvar orifice. The orifice moves anteriorly and apically into the cavity, away from the ventricular inlet, and toward the outflow tract (Fig. 2). In some patients, the right side of the anterior leaflet can also show failure of delamination with rotational displacement, although in these circumstances it can be difficult to differentiate between the anterior and inferior leafets.

The hinge points of the partially delaminated leaflets also rotate around the aortic root, resulting in the effective valvar orifice closing at the junction of the inlet and apical trabecular portions of the right ventricle. This is in contrast to the normal level of closure, which is at the atrioventricular junction. Because of this change, there is usually tricuspid regurgitation, although rarely the abnormal valve may close in competent fashion. The tricuspid leaflets themselves are typically bizarre and dysplastic, and are tethered by short cords and papillary muscles, or are attached to the underlying myocardium directly or by muscular bands. The tendinous cords may be few in number or absent. Fenestrations of the leaflets are common. These are characteristically comprised of an opening in the leaflet guarded by a single papillary muscle, which gives origin to cords which attach around the periphery of the opening. The result of all of these abnormalities is that the leaflets typically meet as pouches, instead of coapting in competent fashion.

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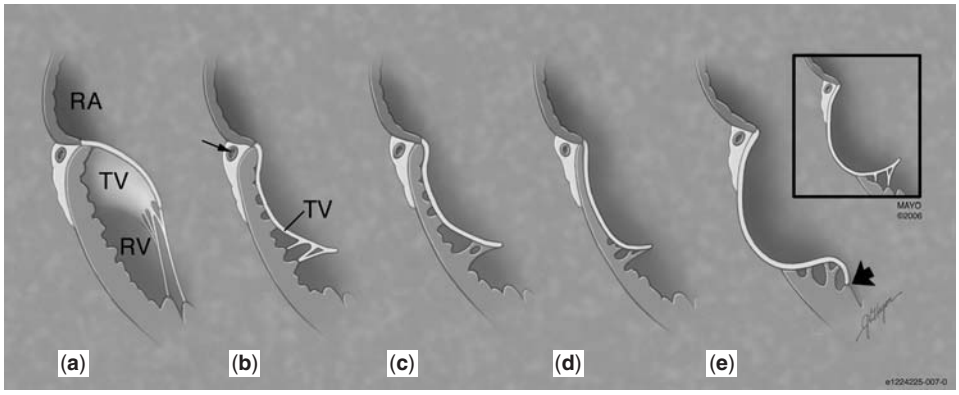


**Figure 1.**
*Coronal view of the process of delamination of the tricuspid valve. There is an infinite anatomical variability when failure of delamination occurs so that no two hearts with Ebstein's malformation are alike.* (***a***)*, Normal tricuspid valve with complete delamination. Leaflets are not adherent to the underlying normal myocardium, the leading edge is free, and the cordal support is normal. RA, right atrium; RV, right ventricle; TV, tricuspid valve.* (***b***)*,* (***c***)*,* (***d***)*, Progressive failure of delamination of the leaflets. Adherence of the leaflet tissue to the underlying abnormal myocardium. The leading edge may have areas of attachment to the underlying myocardium. The area between the true and functional tricuspid annulus is the atrialized portion of the right ventricle. The arrow points to the right coronary artery.* (***e***)*, Nearly complete failure of delamination, with a markedly thinned area of the atrialized right ventricle. The leading edge in this example has direct attachments to the underlying myocardium (arrowhead).* Inset, *This demonstrates an example of nearly complete failure of delamination in one area of the leaflet, with a thinned atrialized ventricle, but with the presence of a free leading edge of the leaflet.*

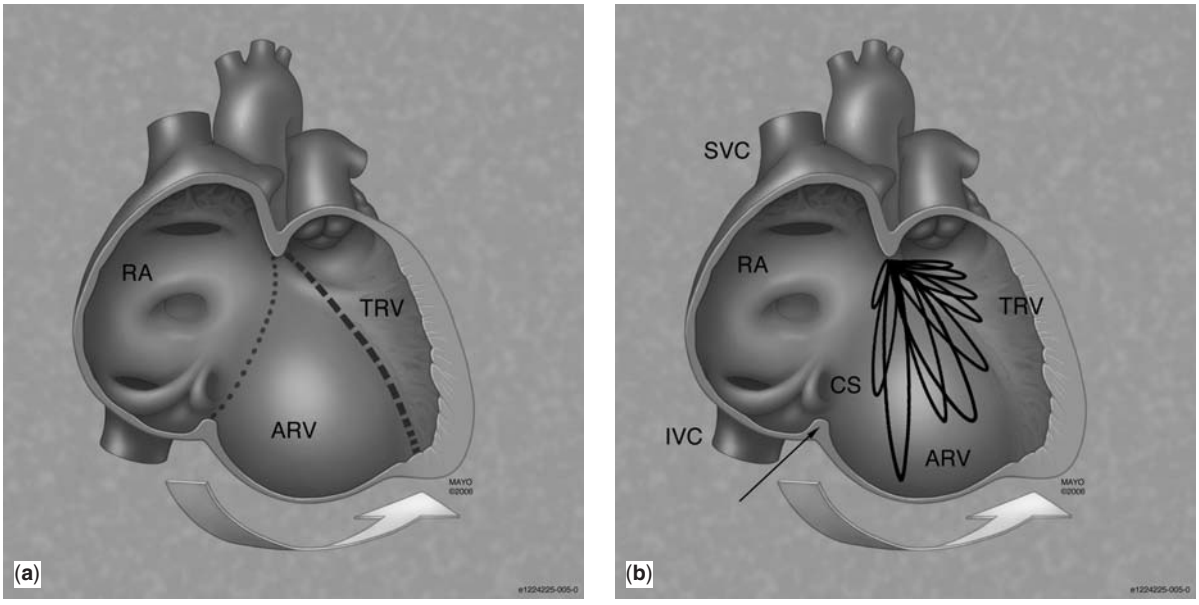


**Figure 2.**
(***a***)*, Normal proximal tricuspid attachments at the atrioventricular junction (circle dotted line) and the direction of the hinge line in Ebstein's malformation (square dotted line). The displacement of the orifice of the valve is* rotational *(flat arrow). ARV, atrialized right ventricle; RA, right atrium; TRV, true right ventricle.* (***b***)*, The location of the functional orifice of the abnormal valve (black ovals) as observed in the series of hearts examined by Schreiber et al.*[15] *Abbreviations: CS: coronary sinus; IVC: inferior caval vein; SVC: superior caval vein.*

In the most anatomically severe cases, the septal leaflet is often no more than a ridge of fibrous tissue that originates below the membranous septum and is directed toward the apex. No other remnant of the septal leaflet is present. In addition, there may be little evidence of delamination of the inferior leaflet; with only a few remnants of leaflet tissue or thickened endocardium seen towards the ventricular apex. The anterior leaflet may also be severely deformed so that the only mobile leaflet tissue present, often unsupported by any tendinous cords, is displaced into the right ventricular outflow tract (Fig. 2b).

A characteristic of the anterior leaflet that is critical to most attempted repairs, but one that does not always

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receive due attention, is the presence of a free leading edge. The leading edge of the anterior leaflet can be free and mobile, or it may have hyphenated or linear direct attachments to the underlying endocardium. In the most severe cases, the linear attachment is complete, the leading edge being directly attached to the endocardium throughout its length.[1] In each case, nonetheless, there can be partial or complete delamination of the remaining portion of the leaflet (Fig. 1, inset). The presence of a free leading edge increases the probability of obtaining a successful and durable repair, should reconstruction be attempted.

There is also variation in the degree of rotational displacement of the functional from the true annulus. As already emphasized, the true annulus remains in its normal location at the right atrioventricular junction, albeit that it may be poorly defined, especially posterolaterally, and is usually dilated. The right ventricular wall separating the true from the functional annulus is referred to as the "atrialized" right ventricle. This atrialized right ventricle is typically dilated, and often thinner than normal, with the most extreme thinning being found inferiorly (Fig. 3). Dilation of the right ventricle may be massive. It involves not only the right ventricular wall proximal to the tricuspid valve, but also the right ventricle distal to the valve, this being the so-called functional right ventricle, which includes the right ventricular infundibulum. These findings suggest that the cardiac dilation is due not only to the haemodynamic abnormalities of the anomaly, but also to a generalized right ventricular myocardial dysfunction. Morphometric histopathologic studies have demonstrated that right ventricular dilation is associated not only with thinning of the wall, but also with an absolute decrease in the number of myocardial aggregates counted through the thickness of the wall from endocardium to epicardium.[2] Interstitial fibrosis may also develop in chronic cases.

Marked dilation of the right ventricle significantly affects the structure and function of the ventricular septum and left ventricle. The left ventricle is compressed, displaced posteriorly, and rotated toward the spine, an arrangement also described as "pancaking". In the more severe cases, the septum is flattened, or shows D-shaped leftward bowing. This can be associated with paradoxical septal motion (Fig. 4). Although measurements of fractional shortening and ejection fraction by echocardiography are less reliable in the presence of paradoxical motion of the septum, many such patients clearly have depressed left ventricular systolic function and a reduced ejection fraction.

## Repair of the tricuspid valve

Repair is based predominantly on a satisfactory arrangement of the anterior leaflet. Thus, significant abnormalities of the leaflet may compromise the result. Fenestrations or perforations in the anterior leaflet can usually be repaired satisfactorily with fine running or interrupted sutures. It may be possible to construct a competent valve when the anterior leaflet is small, but this will likely be at the possible expense of creating some degree of tricuspid stenosis, which is nearly

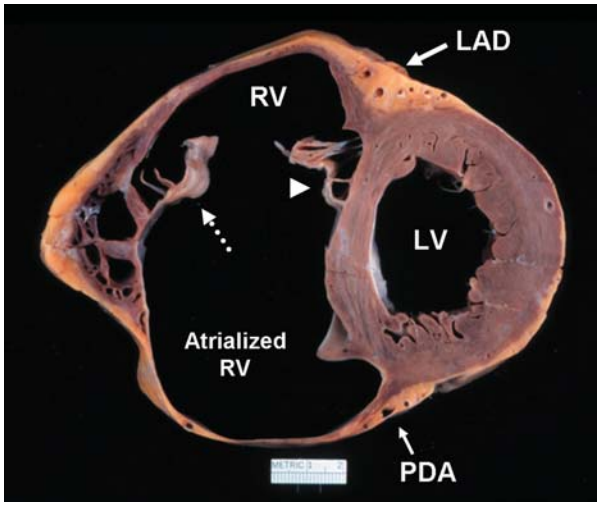


**Figure 3.**
*Horizontal cross-sectional view at the midventricular level of a heart with Ebstein's malformation. Arrowhead, septal tricuspid valve leaflet; dotted arrow, anterior tricuspid valve leaflet; LAD, left anterior descending coronary artery; LV, left ventricle; PDA, posterior descending coronary artery; RV, right ventricle.* (Photo courtesy of William D. Edwards.)

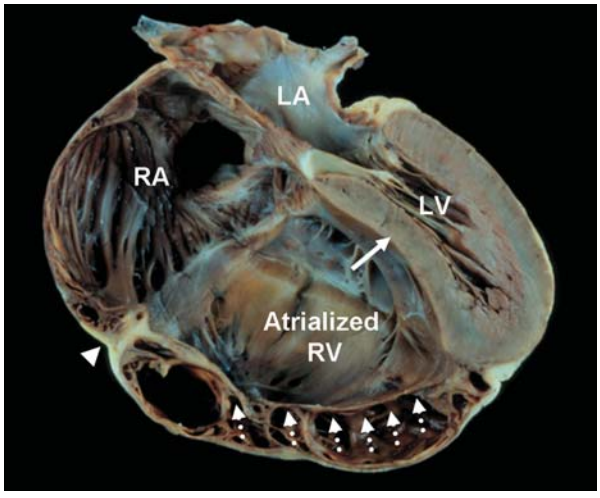


**Figure 4.**
*Four chamber cross-sectional view of a heart with Ebstein's malformation. Arrowhead, anatomic right atrioventricular junction (true tricuspid annulus); dotted arrows are located in the functional right ventricle and point to partially delaminated tricuspid leaflet tissue; solid arrow points to a deviated ventricular septum from the enlarged right ventricle; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.* (Photo courtesy of William D. Edwards.)

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always acceptable. The most important feature for a satisfactory repair is the presence of a mobile and free leading edge of the anterior leaflet. Repair is especially difficult when the anterior leaflet has hyphenated or linear attachments to the underlying endocardium. The presence of short papillary muscles and cords does not preclude a satisfactory repair if the remaining leaflet tissue is well formed. Direct insertion of the heads of the papillary muscles into the leading edge of the anterior leaflet, however, severely restricts its motion, making repair more difficult. Some patients will have a large enough inferior leaflet to permit a bifoliate repair and, rarely, all three leaflets will be moderately well formed but displaced, permitting a trifoliate repair.

Our original repair consisted of plication of the free wall of the atrialized portion of the right ventricle, an inferior annuloplasty, and excision of redundant right atrial wall, in other words a reduction atrioplasty.[3] The repair was based on the construction of a unifoliate valve, using the anterior leaflet, which as explained is usually enlarged in this setting. In fact, numerous techniques have been described for repair of the valve since the first report of Hunter and Lillehei.[4] This is hardly surprising, since as we have explained, no two hearts show exactly the same anatomy, which makes it difficult to employ a standard system for classification.[5,6] While the basic principles of our original repair remain the same, we have incorporated various modifications in the repair as numerous anatomical variants of the anomaly have been encountered and our experience has grown (Figs. 5–7). The original inferior pursestring annuloplasty may be modified by bringing the tricuspid annulus at the appropriate point on the right ventricular free wall directly to the ventricular septum, where it is anchored with pledgeted sutures. The remainder of the inferior annulus is then obliterated with pledgeted mattress or running sutures.

Other modifications we have employed include bringing the intact anterior papillary muscle, or muscles, toward the ventricular septum with one or more

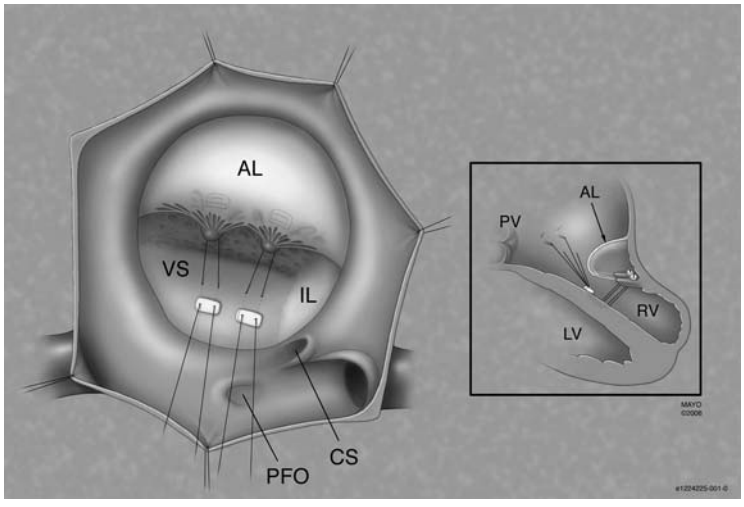


**Figure 5.**
*Basic principles of one of our current technique for repair of the tricuspid valve. The manoeuvres are designed to progressively bring the leading edge of the anterior leaflet (AL) closer to the ventricular septum (VS), or septal leaflet, in order to optimize leaflet coaptation and establish competence of the valve. The base of the intact major papillary muscle(s), which arises from the free wall of the right ventricle, is moved toward the ventricular septum at the appropriate level with a pledgeted horizontal mattress sutures. CS, coronary sinus; IL, inferior leaflet; PFO, patent foramen ovale.* Inset, *Coronal view of the right ventricle (RV) and right atrium demonstrating a small dimple effect that occurs in the anterior free wall of the right ventricle after this manoeuvre is completed. LV, left ventricle; PV, pulmonary valve.*

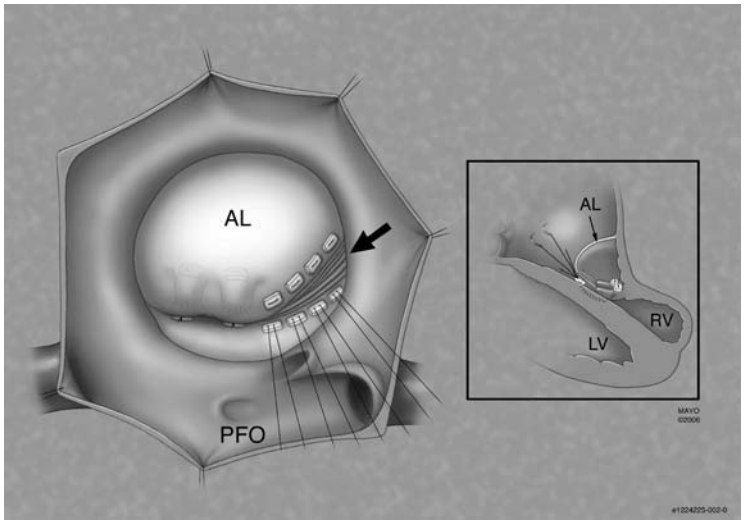


**Figure 6.**
*The inferior angle of the tricuspid orifice is closed by bringing the right side of the anterior leaflet down to the septum and plicating the nonfunctional inferior leaflet in the process (arrow).* Inset, *After all of the mattress sutures are secured, improved proximity of the leading edge of the anterior leaflet with the ventricular septum is noted.*

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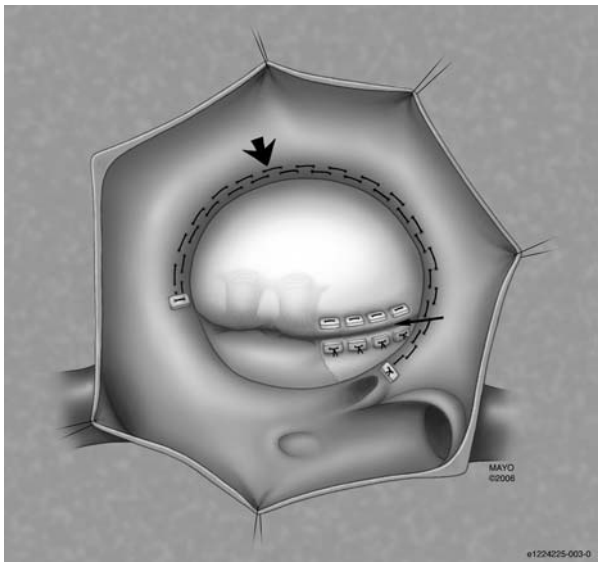


**Figure 7.**
*Plication of the inferior angle of the annulus with pledgeted mattress sutures (arrow). An anterior pursestring annuloplasty (arrowhead) may be performed to further narrow the tricuspid annulus. This annuloplasty may begin at the anteroseptal commissure, anterior to the membranous septum, and ends beyond the inferoseptal commissure, adjacent to the coronary sinus. Alternatively, the annuloplasty can be performed posterolaterally to reduce the size of the annulus, which also brings the free wall closer to the septum.*

pledgeted sutures in order to facilitate closure and contact of the anterior leaflet against the ventricular septum, and the addition of an anterior pursestring annuloplasty to the repair of those hearts that have extensive dilation of the right ventricular outflow tract and anterior portion of the tricuspid annulus. It is occasionally the case that the valvar morphology is adequate for a bifoliate or trifoliate repair. This is usually accomplished with an annuloplasty at the level of the displaced annulus.

The original technique of plicating the ventricular wall from apex to base results in maximal reduction of right ventricular size, but this approach appeared possibly to be associated with increased disturbances of ventricular rhythm. Currently, we reserve plication or resection for patients having thin, transparent, or dyskinetic segments of the atrialized right ventricular myocardium. These changes are most often seen in the area of the inferior wall of the right ventricle (Fig. 8). Potential advantages of plication or resection include:

- reduction in the size of the nonfunctional portion of the right ventricle, which speeds transit of blood flow through the right heart,
- reduction of compression of the left ventricle by the pancake effect, thus improving left ventricular function,
- elevating the papillary muscles, which facilitates closure of the anterior leaflet against the septum in systole, and
- providing more space for the lungs, this being especially important in infants.

Disadvantages of right ventricular plication include potential compromise of the coronary arterial supply to right ventricular musculature, and potential development of ventricular arrhythmias.

## Replacement of the tricuspid valve

In general, we believe repair is preferable to replacement whenever repair is feasible. If there is failure of delamination of more than half of the anterior leaflet, or if the leading edge of the anterior leaflet has hyphenated or linear attachment to the right ventricle, a durable repair may not be obtainable with techniques of valvoplasty. In these circumstances, replacement is then preferred. Replacement should be performed in a manner that protects the conduction tissue and the right coronary artery.[5,6]

When replacement is performed, it is important to resect any leaflet tissue which is displaced toward the right ventricular outflow tract, since this can obstruct the right ventricular outflow tract. A prosthetic valve, bioprosthetic more often than mechanical, is then inserted (Fig. 9). The suture line is deviated to the atrial side of the atrioventricular node and membranous septum to avoid injury to the conduction mechanism. A small vein crossing the tricuspid annulus adjacent to the membranous septum typically marks the site of the atrioventricular node. In order to avoid injury to the right coronary artery, the suture line is deviated cephalad to the true tricuspid valve annulus inferolaterally, where the tissues are frequently very thin. The coronary sinus can be left to drain into the right atrium if there is sufficient room between it and the atrioventricular node. If the distance is short, then the coronary sinus can be left to drain into the right ventricle. In our experience, the drainage postoperatively of the coronary sinus has not been noted to affect left ventricular function.

## The role of the bidirectional cavopulmonary shunt

In our experience, the overwhelming majority of patients with Ebstein's malformation can undergo successful biventricular repair. We currently use the bidirectional cavopulmonary shunt on a selective basis, typically in the setting of a severely enlarged and/or severely dysfunctional right ventricle as part of the one-and-a-half ventricle repair. Because concomitant

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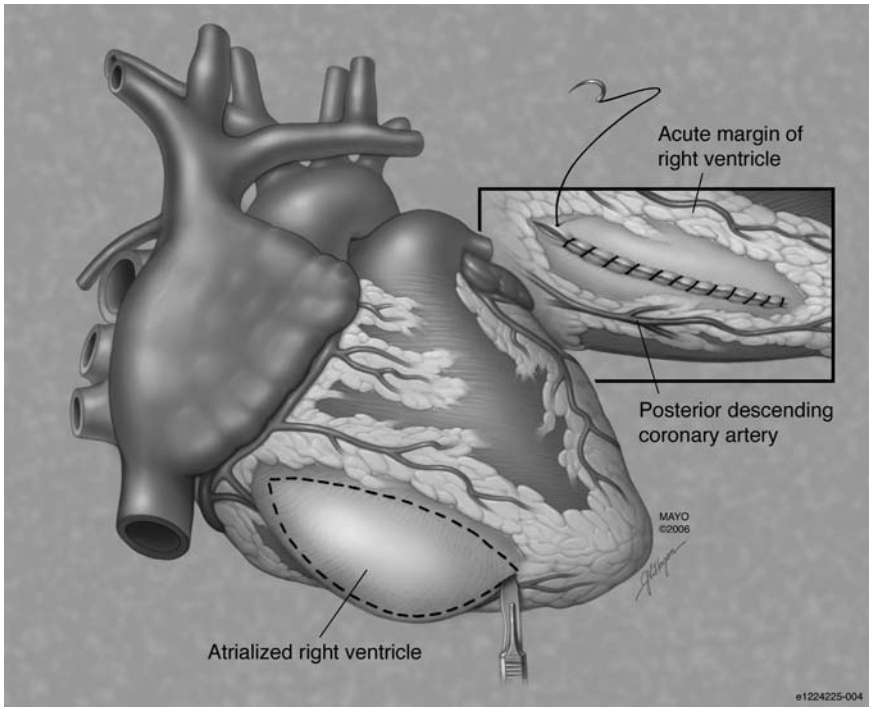


**Figure 8.**
*Resection of a markedly thinned atrialized portion of the right ventricle, usually the inferior wall. The resection (or plication) is performed from the base toward the apex and parallel to the coronary arteries.* Inset, *Completed suture line is parallel to the long axis of the heart. The acute margin of the right ventricle is effectively brought close to the posterior descending coronary artery.*

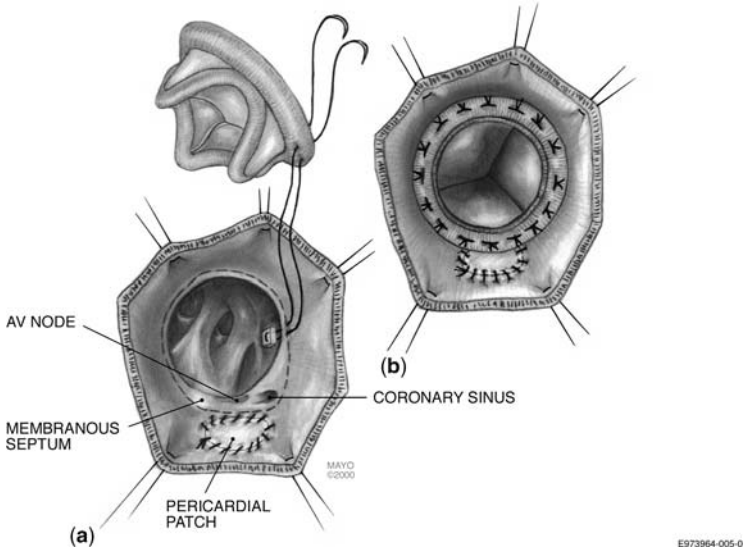


**Figure 9.**
*Diagram of technique for replacement of the tricuspid valve in Ebstein's malformation. (**a**), The valve suture line is placed on the atrial side of the membranous septum and atrioventricular node (AV) to avoid injury of the conduction system. The struts of the bioprosthetic valve are positioned so that they straddle the conduction system in order to avoid injury to it. 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. When there is sufficient distance between the coronary sinus and the AV node, the coronary sinus may be left on the atrial side of the suture line. (**b**), The sutures are tied with the heart perfused and beating to ensure that the conducted rhythm is preserved.*

left ventricular dysfunction may be present when the right ventricle is massively enlarged, it is important to document by direct measurements that the pulmonary arterial and left atrial pressures are low. This may require a preoperative cardiac catheterization, albeit that direct intraoperative measurements are routine prior to considering construction of the bidirectional cavopulmonary shunt. The shunt accomplishes two important things in the setting of right ventricular failure. First, it reduces the venous return to the enlarged and dysfunctional right ventricle by almost half. Second, it optimizes preload to the left ventricle.

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Construction of the shunt is permissible in the setting of depressed left ventricular function provided the left ventricular end-diastolic pressure is less than 15 millimetres of mercury, the transpulmonary gradient is less than 10 millimetres of mercury, and the mean pulmonary arterial pressure is less than 18 to 20 millimetres of mercury. The shunt is not constructed on a routine basis because it compromises subsequent access to the heart for electrophysiological evaluation or therapeutic manoeuvres such as ablation or placement of pacemaker leads. It also has late potential complications, such as pulsations in the neck veins and head, facial suffusion, and development of collateral veins and pulmonary arteriovenous fistulas.

Should a shunt be performed concomitantly at the time of replacement of the tricuspid valve, the flow of blood through the tricuspid valve is decreased. It is important, therefore, to place a valve that is not oversized. In other words, a valve that matches the reduced flow through the right heart, so that all the leaflets of a bioprosthesis, or the discs of a mechanical valve, open and close normally.

## Management of supraventricular tachyarrhythmias

The pathologic abnormalities in Ebstein's malformation, in addition to marked enlargement of the right atrium and right ventricle, include an increased incidence of associated accessory muscular pathways for atrioventricular conduction, which provide the substrate for the development of both supraventricular and ventricular tachyarrhythmias. These arrhythmias are usually poorly tolerated. Although current surgical procedures of valvar repair or replacement, with correction of associated defects, are highly successful, and yield excellent early and late results, long-term outcome can still importantly be affected by the state of cardiac rhythm. Many of the atrial tachyarrhythmias can be managed preoperatively by catheter techniques, but the success rate is lower than for patients who do not have Ebstein's malformation for several reasons, including distortion of the anatomic landmarks, presence of tricuspid regurgitation, catheter instability, tendency toward development of atrial fibrillation during catheter manipulation, and the presence of multiple broad bands of accessory muscular pathways rather than the discrete pathways typically found in the setting of Wolff-Parkinson White syndrome in the absence of Ebstein's malformation. Because the rate of late success for catheter ablation of accessory pathways is suboptimal, we favour primary reliance on surgical ablation.

We advocate preoperative electrophysiologic testing for all patients with Ebstein's malformation who have evidence of preexcitation by electrocardiography, or who have a history of wide-complex tachycardia or tachyarrhythmia of unknown type. Electrophysiologic testing can help to identify the presence, number, and characteristics of accessory pathways as well as concealed pathways, atrioventricular nodal reentrant tachycardia, and easily-inducible atrial flutter and fibrillation. Because surgical procedures are now available to treat these arrhythmias, their proper identification before corrective surgery is important for optimal control of rhythm after the operation.

At the time of repair of the malformation, concomitant electrophysiologic mapping is performed for patients with accessory pathways and the pathways are ablated by surgical division and/or cryoablation. Atrioventricular nodal reentry tachycardia is treated by concomitant perinodal cryoablation, and atrial flutter and fibrillation are treated by a concomitant right-sided maze procedure.[6–10] In our experience, the addition of one or more concomitant procedures aimed at preventing arrhythmias at the time of corrective surgery did not increase the early mortality, at 4.8 percent, relative to the overall mortality rate of 6.8 percent for the overall series of 498 patients.[11] At late follow-up, there was complete freedom from recurrent atrioventricular nodal reentry tachycardia, and reciprocating tachycardia mediated by accessory pathways. Freedom from one or more episodes of recurrent atrioventricular atrial flutter or fibrillation after a right-sided maze procedure was 75 percent, but only 7 percent of late survivors still had an arrhythmia after adjunctive medical and/or catheter therapy.[11,12]

## Results for surgery at the Mayo Clinic

Between April 18, 1972, and April 1, 2006, operations have been performed on patients admitted to the surgical services controlled by the authors in 551 consecutive patients with concordant atrioventricular and ventriculoarterial connections and biventricular circulations. The ages at operation ranged from 2 months to 79 years, with a median of 21 years and a mean of 24 years. Repair was performed in 189 patients, and the valve was replaced, usually with a bioprosthesis, in 355 patients. A one-and-a-half ventricle repair was performed in 16 patients (2.9 percent), and the Fontan circulation was created in 3 (0.5 percent). Overall, there were 27 early deaths (4.9 percent). Since 1999, the early mortality has decreased to 2.4 percent in 167 patients. Concomitant procedures included a right-sided maze procedure in 53 patients, ablation of accessory pathways for atrioventricular conduction in 45 patients, and ablation of atrioventricular nodal reentry tachycardia in 8 patients.

Late results of the first 323 patients with Ebstein's anomaly who underwent surgical treatment have

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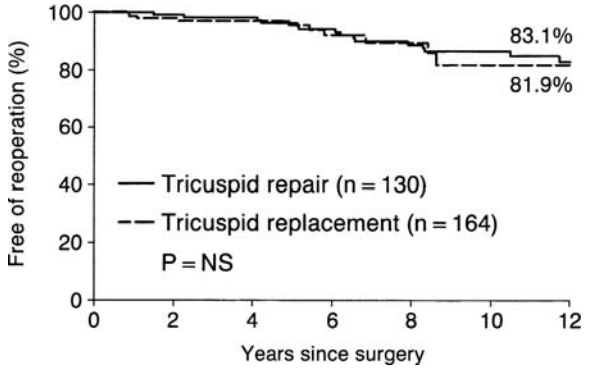


**Figure 10.**
*Freedom from reoperation after replacement of the valve, both bioprosthetic and mechanical, compared with freedom from reoperation after repair. (From Kiziltan HT, Theodoro DA, Warnes CA, et al. Ann Thorac Surg 1998; 66: 1539.)*[5]

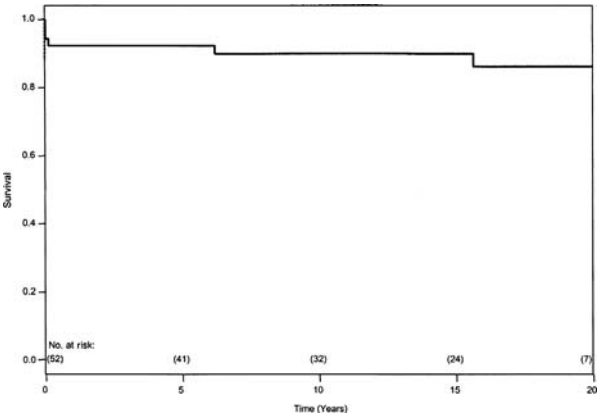


**Figure 11.**
*Kaplan-Meier survival estimates after repair in young children aged less than 12 years. (From Boston US, Dearani JA, O'Leary PW, et al. Ann Thorac Surg 2006; 81: 694.)*[8]

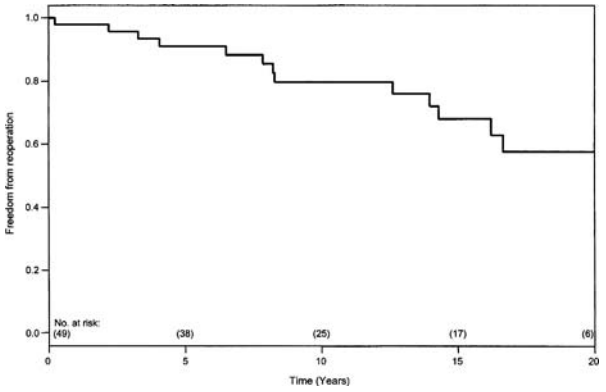


**Figure 12.**
*Freedom from late reoperation for recurrent tricuspid valve regurgitation in young children with Ebstein's malformation. (From Boston US, Dearani JA, O'Leary PW, et al. Ann Thorac Surg 2006; 81: 693.)*[8]

been reviewed.[12,13] There were 23 late deaths (7.6 percent) in a follow-up extending up to 25 years (mean 7.1 years). Late echocardiography showed the atrial septum was intact in all patients. Of the 138 patients who underwent valvar repair, 23 (16.7 percent) subsequently required reoperation from 1.5 to 17.7 years later, with a mean of 9.4 years.

Interestingly, freedom from reoperation at 10 and 15 years after replacement of the valve, using either a bioprosthetic or mechanical substitute, was not significantly different from valvar repair, at 81.9 percent as opposed to 83.1 percent[5] (Fig. 10). Results of repair have recently been reported in 52 young children.[8] Actuarial survival at 5, 10, and 15 years was 92 percent, 90 percent, and 90 percent, respectively (Fig. 11). Freedom from reoperation for recurrent tricuspid regurgitation at 5, 10, and 15 years was 91 percent, 80 percent, and 68 percent, respectively (Fig. 12). Moderate or more severe tricuspid regurgitation as observed on the echocardiogram at the time of dismissal from hospital was the only risk factor for reoperation ( p equals 0.04).

Symptoms were improved postoperatively in the majority of patients, with over nine-tenths being in the first or second classes of the system devised by the New York Heart Association. Postoperative reduction in cardiac size was usual, and occasionally considerable. We know of 9 female patients who have undergone a total of 12 successful pregnancies with delivery of normal children. Maximum exercise testing showed a significant increase in work performance, exercise duration, and maximum uptake of oxygen after the operation.[14] Maximal consumption of oxygen increased from a mean of 47 percent of predicted value before operation to a mean of 72 percent at follow-up. Repair of Ebstein's malformation also favourably affects cardiac output, particularly in response to exercise, normalizes systemic arterial saturations of oxygen, and reduces excess ventilation at rest and during exercise.

## Conclusions

Repair of Ebstein's malformation eliminates right-to-left intracardiac shunting with its attendant risks, improves exercise tolerance and functional class, and reduces supraventricular tachyarrhythmias. In addition, quality of life and longevity are improved.

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