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Surgical Management of Ebstein's Anomaly

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Ebstein's malformation is a congenital anomaly of the tricuspid valve and right ventricle. Surgical repair of Ebstein's anomaly improves functional class and exercise tolerance, eliminates right-to-left intracardiac shunting (if present), and reduces the incidence of supraventricular tachyarrhythmias. As a result, quality of life and survival are improved. Because of the variable degree of malformation present, repair is predicated on favorable anatomic factors, most importantly the arrangement of the anterior leaflet of the tricuspid valve. When anatomic derangements threaten a durable tricuspid valve repair, valve replacement with protection of the conduction tissue and right coronary artery should be performed. The vast majority of patients can undergo a biventricular repair. The application of the bidirectional cavopulmonary anastomosis is reserved for patients with poor right ventricular function. Freedom from reoperation after tricuspid valve repair is similar when compared with valve replacement. In the current era, overall early mortality after surgical repair in children and adults has fallen to less than 3% in experienced centers. Surgical treatment of the symptomatic neonate remains a significant challenge, with approaches that include either a biventricular or single ventricle algorithm.
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Ebstein's anomaly (EA) is a congenital cardiac malformation primarily involving the right ventricle (RV) and tricuspid valve, in which the tricuspid valve leaflets remain variably adherent to the underlying RV myocardium. This failure of delamination results in the characteristic rotational and apical displacement of the tricuspid valve. The extent of delamination and dysplasia of the tricuspid valve is different in each patient, resulting in infinite degrees of anatomic variability. Significant tricuspid regurgitation leads to progressive right atrial and RV dilatation. Atrial and ventricular arrhythmias may coexist. Despite often significant anatomic abnormalities, symptoms may be minimal or absent, especially when there is no (or very small) atrial septal defect present. The timing and choice of treatment depends on numerous factors, including the patient's clinical presentation, morphology of the tricuspid valve, presence of right-to-left shunting through an atrial septal defect, severity of right-sided chamber dilatation, severity of RV dysfunction, or presence of atrial or ventricular arrhythmias.

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Anatomic and Surgical Considerations

The anatomy of the tricuspid valve and RV in EA is characterized by several features that can exhibit a spectrum of malformation. The first is adherence of the tricuspid valve leaflets to the underlying myocardium (ie, failure of delamination). Progressive severity of the malformation results in valve leaflets that are increasingly adherent to the RV myocardium. The septal leaflet is the most severely affected, inferior (posterior) leaflet is less severely involved, and the anterior leaflet is least affected. In the most severe cases, the septal leaflet represents just a ridge of fibrous tissue with no other remnant of normal leaflet present. In addition, the inferior leaflet may only have a diminutive remnant of leaflet tissue present, leaving the anterior leaflet as the only contributor of mobile leaflet tissue. The second feature is anterior and apical rotational displacement of the functional tricuspid annulus. This migration of the functional annulus is into the RV cavity toward the RV outflow tract and away from its normal position at the atrioventricular (AV) junction. The third feature is tethering, redundancy, and fenestrations of the anterior leaflet. Chordae may be foreshortened and bizarre papillary muscle(s) configurations may be present. The fourth abnormality is dilation of the "atrialized" portion of the

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RV, with variable degrees of thinning and hypertrophy of the RV wall. The fifth feature is dilation of the right AV junction (true tricuspid annulus). The final abnormality is the presence of variable ventricular myocardial dysfunction. Importantly, each heart with EA is different, with an infinite variability of the above-mentioned characteristics. Consequently, classification systems for the anatomic variants of EA are difficult.[1,2]

A critical factor to be considered when attempting repair of the tricuspid valve is whether or not there is presence of a free leading edge of the anterior leaflet. This too can be variable, ranging from a freely mobile leaflet edge, to hyphenated attachments, to one that is completely attached (linearly attached) throughout its course.[3] The presence of a mobile leading edge increases and optimizes the likelihood of performing a successful and long-lasting repair.

In addition to leaflet abnormalities, there is also variation in the degree of rotational displacement of the functional tricuspid annulus away from the true annulus. The RV wall separating the true and functional tricuspid annulus is referred to as the "atrialized" portion of the RV. The atrialized RV is typically dilated and thinner than normal, especially inferiorly. Anatomically, the degree of abnormality of the atrialized RV is also quite variable, ranging from minor degrees of myocardial thinning to a large, dyskinetic, severely thinned, and transparent atrialized RV wall. In addition, the size of the functional RV is variable depending on the degree of displacement of the functional tricuspid annulus toward the RV outflow tract. RV dilatation can adversely affect the structure and function of the interventricular septum and left ventricle (LV). The LV may be compressed and displaced posterior, a term that is referred to as "pancaking." Paradoxical septal motion can be present in more severe cases of EA resulting in a decreased ejection fraction and depressed LV systolic function.

Indications for Operation

Patients generally considered for operation are those with progressive symptoms or decreased exercise tolerance, increasing cyanosis, or after occurrence of a paradoxical embolism. In addition, operation is usually advised with increasing cardiomegaly on chest x-ray, worsening RV dilatation or deteriorating RV systolic function by echocardiography, or onset or progression of atrial or ventricular arrhythmias.

Most patients in functional New York Heart Association (NYHA) class I or II with mild or no cardiomegaly can be managed medically. Once symptoms develop and the patient progresses to functional NYHA class III or IV, medical therapy typically becomes ineffective. In borderline situations, if anatomic features favorable for valve repair are observed on echocardiography, then operation is advised earlier. A biventricular repair is usually possible for the vast majority of patients, a 1.5 ventricle repair (concomitant bidirectional cavopulmonary anastomosis [BDCPA]) is used when there is poor RV function, and cardiac transplantation is reserved for patients with severe LV dysfunction.

Surgical Treatment

The surgical approach to EA has continued to evolve over the last 40 years, since the initial report of attempted repositioning of the septal and posterior valve leaflets by Hunter and Lillehei in 1958.[4] Since that time numerous techniques of tricuspid repair have been reported.[2,5–24] The variety of surgical techniques for valve repair emphasizes the enormous anatomic variability that exists.

In our experience, the critical feature necessary for a successful and durable repair is the presence of a mobile and free leading edge of the anterior leaflet. As such, significant abnormalities of the anterior leaflet may compromise the result. Repair is especially challenging if there are extensive hyphenated or linear attachments of the anterior leaflet to the underlying myocardium. If the anterior leaflet tissue is well formed (ie, $>$ 50% delaminated), a successful repair may still be possible even in the presence of short papillary muscles and chordae tendineae provided the leading edge is free and mobile. At the less severe end of the spectrum, there may be enough inferior or septal leaflet to allow a bileaflet repair, and rarely, all three leaflets will be formed enough to permit a trileaflet repair. In general, it is easier to repair and obtain a competent valve when there are two or three well-formed leaflets.

Our original repair, developed by Danielson et al,[21] consisted of transverse plication of the atrialized RV (bringing the apex closer to the base), posterior tricuspid annuloplasty, and right reduction atrioplasty. This repair was based on the construction of a functional "monocusp" valve based on satisfactory anatomy of the anterior leaflet. The posterior annuloplasty optimizes coaptation of the anterior leaflet with the ventricular septum. The transverse plication effectively brings the functional tricuspid annulus up toward the true annulus. While this technique results in a marked reduction in the size of the RV, its main disadvantage is potential compromise of branches of the right coronary artery, which run perpendicular to the direction of plication.

Consequently, as our experience has grown to more than 500 consecutive surgical cases, we have incorporated modifications to our original repair to address the numerous anatomic variations of the malformation.[22–24] The techniques currently used focus on repair of the valve where it exists in the RV (ie, repair at the level of the functional annulus). Modifications include moving the base of the intact papillary muscle on the RV free wall toward the ventricular septum at the appropriate level with interrupted, pledgeted mattress sutures (Fig. 1). It is important to place these mattress sutures deep into the ventricular septum. The defect at the inferior tricuspid annulus is obliterated by approximating the right side of the anterior leaflet and annulus down to the ventricular septum, thus improving proximity of the leading edge of the anterior leaflet with the ventricular septum (Fig. 2). When inferior leaflet tissue can be absent, this area is often the site of residual regurgitation. A pursestring annuloplasty can be performed to further narrow the tricuspid annulus, especially in patients with extensive annular dilatation (Fig. 3). This can begin at the anteroseptal commissure and end at

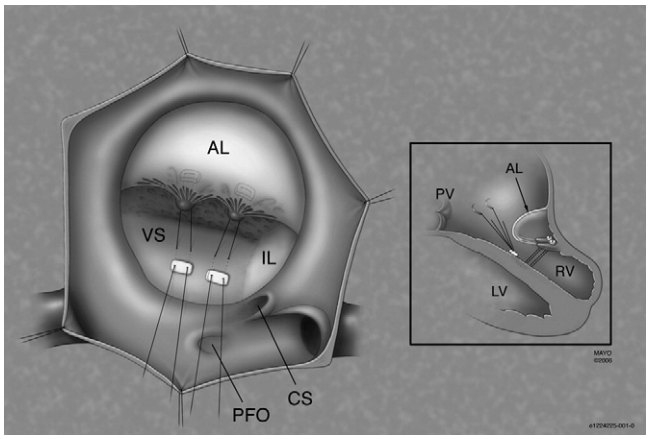

Figure 1 Basic principles of one of our current techniques for repair of the tricuspid valve. The maneuvers are designed to progressively bring the leading edge of the anterior leaflet (AL) closer to the ventricular septum (VS), or septal leaflet to optimize leaflet coaptation and establish competence of the valve. The base of the intact major papillary muscle, which arises from the free wall of the right ventricle, is moved toward the ventricular septum at the appropriate level with 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 maneuver is completed. LV, left ventricle; PV, pulmonary valve. (Reprinted with permission.[24])

the inferoseptal commissure (adjacent to the coronary sinus), or can be performed inferolaterally only. When there is some delamination of the septal leaflet, albeit displaced inferiorly, the addition of an Alfieri stitch, thus creating a double orifice tricuspid valve, can be added to the above maneuvers to optimize coaptation and improve valve competency. In our experience, each tricuspid valve repair is slightly different, and successful repair is best accomplished by incorporation

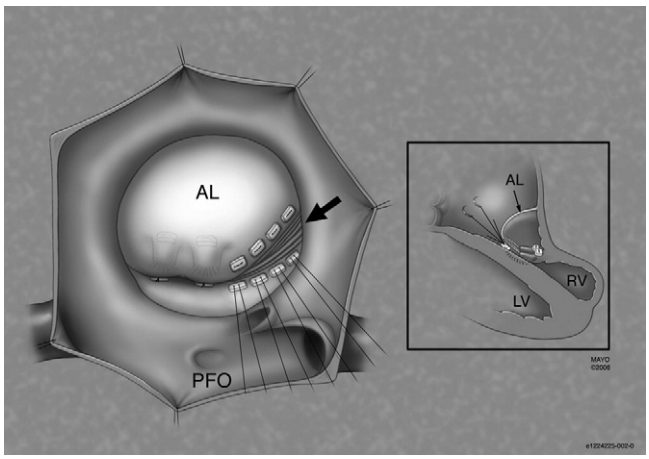

Figure 2 The inferior angle of the tricuspid orifice is closed by bringing the right side of the anterior leaflet down to the septum and plicating the nonfunctional inferior leaflet in the process (arrow). Inset, After all of the mattress sutures are secured, improved proximity of the leading edge of the anterior leaflet with the ventricular septum is noted. (Reprinted with permission.[24])

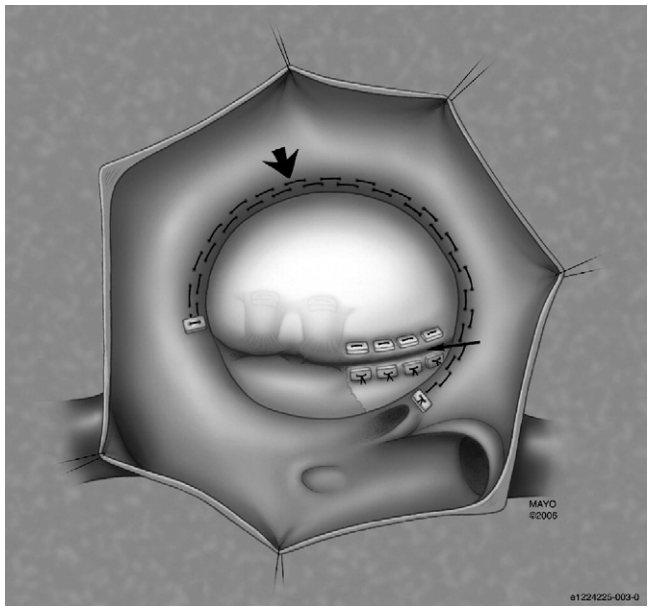

Figure 3 Plication of the inferior angle of the annulus with pledgeted mattress sutures (arrow). An anterior pursestring annuloplasty (arrowhead) may be performed to further narrow the tricuspid annulus. This annuloplasty may begin at the anteroseptal commissure, anterior to the membranous septum, and end 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. (Reprinted with permission.[24])

of the appropriate techniques depending on the anatomy encountered at the time of operation.

Plication or resection of the atrialized RV is performed selectively. We now prefer a longitudinal resection for a large area(s) of a thinned, transparent, and dyskinetic atrialized RV, usually the inferior wall.[24] The resection is usually performed between the acute margin of the RV and the posterior descending coronary artery (ie, inferior wall of the RV). This elliptical resection is parallel to the acute marginal branches of the right coronary artery, thus minimizing coronary arterial compromise. Resection of both the anterior and inferior RV walls has also been described.[25] Decreasing the size of the RV with resection techniques may also reduce LV compression (thus improving LV systolic function) can improve proximity of the anterior leaflet with the ventricular septum, and can offer more area in the chest for the lungs (especially important in infants). Plication is used when there are small areas of thinned, transparent atrialized RV.

We prefer valve repair to replacement when favorable anatomy is present and allows a competent and durable result. However, we recognize the wide variability present with the malformation, and ideal anatomy for a durable repair may not be possible when there is insufficient delamination of the anterior leaflet, or there is extensive linear attachment or multiple hyphenated attachments of the leading edge to the underlying myocardium. In these circumstances, tricuspid valve replacement should be considered, paying special attention to preservation of the conduction tissue and avoiding

injury to 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 tricuspid valve, we resect the displaced anterior leaflet tissue toward the RV outflow tract to avoid potential RV outflow tract obstruction after tricuspid valve replacement. A porcine bioprosthesis is used most often because of excellent durability in the tricuspid position and the ability to avoid long-term warfarin anticoagulation.[26] In the tricuspid position, we prefer a porcine over a pericardial bioprosthesis because of the relatively stiffer leaflets present in a pericardial prosthesis and the typically low right atrial and low RV pressures present in EA. A mechanical prosthesis is used when warfarin anticoagulation is required for another reason. During insertion of the prosthesis, 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). To avoid injury to the right coronary artery, 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 their struts straddle the membranous septum and conduction tissue. The valve sutures are tied while the heart is perfused and 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. We have recently analyzed the results of right atrial versus RV drainage of the coronary sinus in patients with EA and noted no significant difference in LV function at late follow-up. Details of this patient subgroup will be forthcoming in a separate publication.

Role of the 'One and a Half Ventricle Repair'

In EA, the BDCPA, or "one and one-half (1.5) ventricle repair" is used when the RV is judged not capable of supporting the pulmonary circulation.[27,28] The diversion of superior vena caval blood to the pulmonary arteries decreases the volume load on the RV by approximately one half in infants and one third in adults and thereby may decrease RV work.[29] We believe the 1.5 ventricle repair may be an alternative for patients with severe EA and poor RV function who would be at high risk for standard surgical treatment. In patients with EA and poor RV function, the BDCPA achieves two goals: 1) it decreases the volume load of the poorly functioning RV; and 2) provides preload to the LV.

Postoperative RV failure is associated with increased early mortality in surgically treated patients with EA[30] and an enlarged heart is also a risk factor for mortality.[31] Chauvaud et al[30,32] believe that the addition of a BDCPA to the surgical treatment of patients at risk of RV failure decreases operative mortality.

In our experience, the overwhelming majority of patients can undergo a biventricular repair, even in the presence of a significantly dilated RV with moderate RV dysfunction. We have reserved the BDCPA for the more severe cases of RV dysfunction. The BDCPA is not performed routinely because it compromises access to the right heart for electrophysiological evaluation and ablation, and for potential pacemaker lead placement in the future. In addition, the BDCPA can be associated with pulsations of the head and neck veins, facial suffusion, and the development of collateral veins and pulmonary arteriovenous fistulae.[33]

Blood flow through the tricuspid valve is reduced when a concomitant BDCPA is performed at the time of tricuspid valve replacement. Therefore, it is important to place a tricuspid valve prosthesis that is not oversized (ie, a valve that matches the reduced flow through the right heart), so that all the bioprosthetic leaflets or mechanical discs open and close normally. Because we have occasionally noted decreased mobility of the leaflets of bioprosthetic valves after the BDCPA, we now routinely anticoagulate with warfarin for a 3- to 6-month period or until full mobility of all leaflets is demonstrated on echocardiography. Aspirin (81 mg daily) is continued thereafter. The increased risk of thrombosis and need for anticoagulation are also potential disadvantages of a concomitant BDCPA at repair of EA.

When the BDCPA is being considered, it is important to know preoperatively if hemodynamics will allow a BDCPA. In general, elevated left-sided filling pressures or elevated pulmonary artery pressures are uncommon in most patients with EA. However, in the setting of LV dysfunction or when echocardiographic findings suggest elevated left-sided filling pressures or elevated RV systolic pressure, preoperative cardiac catheterization should be performed. In addition, intraoperative direct pressure measurements are obtained before performing a BDCPA. The BDCPA is permissible provided the LV end-diastolic pressure is < 15 mm Hg, the transpulmonary gradient is < 10 mm Hg, and the mean pulmonary artery pressure is < 18 to 20 mm Hg.

Management of Supraventricular Tachyarrhythmias

In addition to the anatomic derangements of marked right atrial and RV dilatation seen in patients with EA, there is also an increased incidence of accessory conduction pathways.[34–36] Each of these independently and collectively provide the anatomic substrate for the development of supraventricular and ventricular arrhythmias, which are poorly tolerated in this patient population that already has a poor physiologic reserve.[34,37–39] Although surgical repair of EA and other congenital heart diseases results in significant improvement in functional capacity, postoperative cardiac arrhythmias continue to be a significant source of morbidity.[38,39]

Preoperative catheter-based intervention is successful for managing many arrhythmias; however, the results in patients with EA is significantly lower compared with those without the condition. This is most likely because of altered anatomy, turbulent flow of significant tricuspid regurgitation, and broad-based accessory muscular pathways instead of the distinct accessory pathways seen in typical Wolff-Parkinson White syndrome without EA.[24,36]

We recommend preoperative electrophysiologic mapping for all patients with EA who have objective evidence of preexcitation by electrocardiography, and for those with a history of tachyarrhythmias. Preoperative electrophysiologic mapping is useful not only to identify accessory conduction pathways, but can also uncover easily inducible atrial flutter and fibrillation. In general we prefer the accessory conduction pathway to be ablated in the catheterization laboratory before surgery when feasible. When percutaneous catheter-based ablation is unsuccessful, then intraoperative mapping and surgical division and cryoablation of the pathway is performed at the time of intracardiac repair.

We use a concomitant right-sided maze procedure during intracardiac repair for patients with EA and documented atrial tachyarrhythmias.[34,36,40,41] While previous studies have shown that the freedom from atrial tachyarrhythmias at late follow-up is modestly reduced by repair of the congenital heart defect, which includes restoring a competent tricuspid valve combined with right reduction atrioplasty,[34,37,40] our experience shows that late results are substantially improved by the addition of a concomitant right-sided maze at the time of intracardiac repair.[35,36,41]

Our current practice is to perform a right-sided Cox-maze III procedure when there is a history of paroxysmal atrial fibrillation or atrial flutter. In addition, when atrial flutter is present, we also perform cryoablation of the right atrial isthmus (inferior vena cava to coronary sinus to tricuspid annulus). When chronic atrial fibrillation is present, we perform a complete biatrial Cox-maze III procedure. Currently, we most often use cryoablation to perform these lesion sets. In our experience, there has been no increase in early morbidity or mortality associated with the addition of a concomitant right-sided maze procedure at the time of repair of congenital heart disease.[35,41]

Various modifications of the Cox-maze procedure used by others for patients with atrial arrhythmias associated with congenital heart disease have also shown beneficial effects.[42,43]

The Mayo Clinic Experience

Overall

Between April 1972 and April 2006, 551 consecutive patients with concordant AV and ventriculoarterial connections and biventricular circulations underwent surgical repair for EA at our institution on the authors' surgical services. Ages ranged from 2 months to 79 years, with a median age of 21 years. Tricuspid valve repair was performed in 189 patients, and valve replacement in 335, usually with a bioprosthesis. A one and a half ventricle repair was performed in 2.9% of the patients, and the Fontan procedure in 0.5%. Overall, early mortality was 5%; however, since 1999, this has decreased to 2.4% (n = 167). A concomitant right-sided maze procedure was performed in 53 patients, surgical division of an accessory conduction pathway in 45, and ablation of AV nodal re-entrant tachycardia in eight. Actuarial survival at 5, 10, and 15 years postoperatively was 92%, 90%, and 90% respectively, during the late follow-up period extending up to 25 years (mean, 7.1 years).[24,26]

Of the first 138 patients who underwent valve repair, approximately 17% required reoperation at a mean time of 9.1 years postoperatively. Interestingly, freedom from reoperation after valve replacement was not statistically different from this.[26] Freedom from reoperation for recurrent tricuspid regurgitation at 5, 10, and 15 years was 91%, 80%, and 68%, respectively, and actuarial overall patient survival at 5, 10, and 15 years was 92%, 90%, and 90%, respectively. Moderate or severe tricuspid regurgitation at hospital dismissal was the only significant risk factor for reoperation ($P = .04$).[24]

Surgical Repair in Young Children

Between October 1974 and November 2003, 52 children under 12 years of age (mean age, 7 years) underwent tricuspid valve repair and annuloplasty.[44] Early mortality was 5.8% prior to 1984; there have been no early deaths since that time (n = 31). Risk factors for early mortality were age < 2.5 years and weight < 10.7 kg. Actuarial survival at 5, 10, and 15 years was 92%, 90%, and 90%, respectively, and freedom from all reoperations at 5, 10, and 15 years was 91%, 77%, and 61%, respectively. Moderate tricuspid regurgitation at hospital dismissal was the only risk factor for reoperation ($P = .04$), and tricuspid stenosis did not occur in any patient. At late follow-up approximately 90% of patients were in NYHA functional class I or II.

Lessons learned from this review emphasize the point that the need for late reoperation is minimized when residual tricuspid regurgitation after repair is mild or less. In addition, despite continued somatic growth, there were no instances of subsequent tricuspid stenosis following valve repair.

Surgical Repair in Older Adults

It is not uncommon for patients with EA to present for operative intervention as older adults. Between April 1972 and December 2004, 60 patients $\geq$ 50 years of age (mean age, 59.6 $\pm$ 7.9 years) underwent operation for EA. Approximately one third of patients were in NYHA functional class III or IV. The majority patients had anatomically severe EA and 25% of patients had evidence of LV dysfunction preoperatively. Early mortality was 5% and LV dysfunction was a risk factor for early death. Five- and 10-year survival was 90% and 75%, respectively. Importantly, advanced preoperative NYHA functional class III and IV and preoperative renal insufficiency (creatinine $>$ 1.3 mg/dL) were risk factors for late death. Eighty-three percent of late survivors were in NYHA functional class I or II at last follow-up.

1.5. Ventricle Repair for EA and the Failing RV

Between July 1999 and March 2006, 14 patients with significant RV dysfunction had a BDCPA as part of their surgical treatment for EA.[45] Three patients were initially evaluated for heart transplantation. The median age at operation was 6 years (range, 17 months to 57.8 years) with all patients having severe anatomic EA of the tricuspid valve and significant RV enlargement and dysfunction. The mean LV ejection fraction was 54.5% (range, 35% to 72%). BDCPA was planned in nine patients and five others underwent the procedure for hemodynamic instability after discontinuation of cardiopulmonary bypass. There was one early death from multisystem organ failure and during follow-up. All patients with preoperative LV ejection $< 50\%$ improved to an ejection fraction $> 50\%$. The 1.5 ventricle repair can be considered as a planned procedure, an intraoperative salvage maneuver, or as an alternative to cardiac transplantation in selected patients.

Management of Supraventricular Tachyarrhythmias

Patients with EA and evidence of right atrial dilatation often have concomitant atrial fibrillation and/or flutter.[34–36,40] From March 1993 to January 2003, 99 patients underwent concomitant right-sided maze procedure during surgical correction of their congenital heart disease.[41] Approximately half of these patients had EA. Median age at operation was 43 years (range, 9 to 72 years) and atrial fibrillation and/or flutter was paroxysmal in greater than 80%. There were six early deaths and four late deaths. At last follow-up (extending up to 8 years), 93% were free from atrial tachyarrhythmias and 82 of the 83 late survivors were in NYHA functional class I or II. A concomitant right-sided maze procedure at the time of intracardiac repair is effective in reducing late recurrent atrial arrhythmias and most patients enjoy an excellent quality of life.

Functional Outcome

Symptoms are improved in the majority of patients, and maximal exercise testing demonstrates increased work performance as well as longer exercise duration. Additionally, repair of EA favorably affects cardiac hemodynamics and normalizes systemic arterial oxygen saturations.[24,46]

Summary

Tricuspid valve repair or replacement is the principal element in the treatment of EA. Concomitant procedures include closure of atrial septal defects, ablation of accessory conduction pathways when present, and maze procedure for atrial tachyarrhythmias. Resection or plication of the atrialized RV is performed selectively. In cases of advanced RV dysfunction, the BDCPA can be applied. Transplantation is reserved for cases of significant LV dysfunction. An individualized approach to the surgical management of EA results in a competent tricuspid valve, eliminates right to left shunting (if present), and reduces atrial tachyarrhythmias. Some patients will require reoperation for recurrent tricuspid regurgitation after tricuspid valve repair or bioprosthetic valve deterioration. Importantly, exercise tolerance, quality of life, and survival are improved for the vast majority of patients.

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