Can We Modify Late Functional Outcome in Ebstein Anomaly by Altering Surgical Strategy?*

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We are once again indebted to the Mayo Clinic congenital cardiac team for a clear, late-outcome study. In this issue of the Journal, Brown et al. (1) evaluate 285 patients with Ebstein anomaly who underwent tricuspid valve surgery (64% of known survivors). There are 3 key findings: 1) patients with Ebstein anomaly who required surgery had a 20-year survival of 76%; 2) the investigators’ perception and some objective data demonstrate a high rate of good functional status; and 3) in addition to tricuspid valve performance, late problems in patients (i.e., long after surgery) suggest that, in patients with Ebstein anomaly, right ventricular performance remains a fundamental problem.

Mechanical Versus Myopathy Issues

Although one tends to think of Ebstein anomaly as a deformity of the tricuspid valve, surgical and echocardiographic inspection of myocardial thickness, and before and after repair function, demonstrate that right ventricular contractility and volume are also abnormal. The tricuspid valve is simply a mechanical problem—one that can be resolved with valve repair or replacement. In the reported dataset, both techniques were equivalent with respect to long-term survival; however, tricuspid valve repair was associated with a greater freedom from tricuspid valve reoperation.

Patients reported their perception of their own exercise capacity. It was perceived to be the same or better than their peers in 49.5% of the cases. Another 31.2% perceived only slightly less exercise tolerance than their peers. The objective observation of a significant number of successful pregnancies indicates that the right ventricle copes reasonably well and contributes to cardiac output reserve. Despite the success of operative therapy on the tricuspid valve, there were findings of symptoms of fatigue (42%), shortness of breath (30%), abnormally rapid heart rate (26%), the need for cardiac medications (66%), and rehospitalization for arrhythmia (21%). Each of these items indicates that cardiac performance is less than optimal. Nevertheless, survival at 20 years (including operative mortality) was 76%, which is an encouragingly high number. Direct comparisons cannot be made, but for perspective, the same group reported that survival for the Rastelli procedure (complex 2 ventricle repair), excluding operative mortality, was 59% at 20 years (2). Single-ventricle Fontan survival at 20 years has been reported to be 60% to 70% (3,4). Cardiac performance appears to be the fundamental issue.

Can We Improve Functional Status by Altering Our Surgical Strategy?

Late-outcome data illuminate the sequelae to operative strategies used many years earlier. This long learning feedback loop allows us to formulate hypothesis regarding potential alterations in surgical strategy that might improve late outcomes. Presenting the heart with a modified physiologic stress may lead to a better long-term cardiac fate.

For example, there is now a broad experience of implanting pulmonary valves for pulmonary insufficiency and dilated right ventricles late after repair of tetralogy of Fallot (TOF). In a look at 36-year outcomes for repaired TOF, Nollert et al. (5) noted that those patients having a greater right ventricle-to-left ventricle pressure ratio appeared to experience improved survival. We have hypothesized that leaving some right ventricular outflow tract stenosis in TOF may be of benefit. Our 16-month post-TOF repair data have demonstrated that anatomically equivalent infants, who had pulmonary “annulus” preservation, had a smaller right ventricle-to-left ventricle pressure ratio compared with those that had a transannular patch (T-J Yun, M. Cheung, G. S. Van Arsdell, unpublished data, 2004). Mild-to-moderate residual obstruction was accepted in the pulmonary annulus preservation group. These findings imply better ventricular health in the short term. However, whether these findings lead to better long-term outcomes will be delineated with time.

The Mayo Clinic data indicate that these patients with Ebstein anomaly fundamentally suffer from less-than-optimal cardiac performance. Do the present data compel us to modify our surgical approach, beyond solving the mechanical issue of tricuspid incompetence, to achieve improved long-term cardiac function? Late risks identified in this article (1) that might be modified by adjunctive surgical work are the development of late atrial arrhythmias and

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possibly the finding that a large right ventricular size was shown to be a contributor to rehospitalization.

**Atrial Arrhythmia**

The development of late atrial arrhythmia has 2 potential etiologies: existing atrial substrate for arrhythmia and subsequent consequence to poor right ventricular performance. In the present study, those patients having atrial reduction plasty were found to have decreased hospital readmissions. It would seem reasonable (as the authors have discussed) to take the next step and hypothesize that, on a cohort basis, a prophylactic right-sided maze would diminish late arrhythmia issues. Alternatively, a subset of patients with very large right atria, greater than a given size, might undergo a prophylactic right-sided maze. Another testable procedure would be to perform uniform atrial reduction along with selective cryoablation to known high-risk areas for reentry circuits. The present study (1) provides data to justify expanding arrhythmia surgery beyond the now standard technique of performing a right-sided maze only for those patients with a history atrial arrhythmia. Routine use of arrhythmia surgery should likely be tested.

The timing of reintervention on a failing tricuspid valve is also important to development of late atrial arrhythmia. Volume load from regurgitation impacts atrial and ventricular health. Stenosis from a failing prosthetic valve can cause atrial arrhythmia. Earlier reintervention, provided that it is safe, may preserve long-term functional cardiac performance, which ultimately is the practitioner’s goal.

**Right Ventricular Size**

Left ventricular failure experience shows that function is improved with a reduction in wall stress achieved via medical and or mechanical means. Leaving the potential medical therapy aside, several mechanical means of improving wall stress for the Ebstein ventricle are available. The elimination of tricuspid insufficiency reduces wall stress and is already standard. Ventricular volume reduction plasty has been performed by some investigators (6,7) (including, more recently, Brown et al. [1]). This technique improves wall stress and may have benefits to the repaired valve, right ventricular systolic function, and left ventricular function through ventricular–ventricular interaction. Ventricular volume reduction is attractive on a theoretic basis. The additional surgical risk appears to be low, but data on its benefits are lacking.

**Ventricular–Ventricular Interaction**

Some cases of severe Ebstein anomaly have been converted to single-ventricle physiology. A radical resection of the right ventricular free wall has been advocated to completely remove volume overloading of the right ventricle. Data on those patients demonstrate restoration of the cylindrical shape to the left ventricle and improved left ventricular contractility (8), which provides further insight into the possibility of improving overall cardiac performance by changing the volume of the right ventricle.

**Cavopulmonary Connection**

Although not uniformly accepted, an additional 25% volume unloading of the ventricle with a bidirectional cavo-pulmonary connection also can be achieved. We and others (including some patients in the report by Brown et al. [1]) have used the addition of a cavo-pulmonary connection in Ebstein’s anomaly, thereby establishing a 1-1/2 ventricle-type repair. Analysis of Ebstein anomaly patients having a 1-1/2 ventricle repair in our institution showed a 10-year survival of 90.2% (9). Interestingly, despite the presence of myopathy, the 1-1/2 ventricle repair Ebstein anomaly patients had better survival at 10 years than those having a 1-1/1 ventricle repair for an anatomically small right ventricle. A further benefit to a cavo-pulmonary connection is the fact that a smaller tricuspid valve annulus can be created, thereby potentially increasing the number of patients that might have repair. A cavo-pulmonary connection can only be used if the pulmonary vascular resistance is low. Whether these wall stress-reducing and potential arrhythmia-reducing adjunctive surgical procedures can be of benefit will need to be determined with further systematic testing and analysis.

**Tricuspid Valve Repair**

Tricuspid valve repair has not been uniformly successful. Durable valve repair is an important contributor to increased time to reoperation or even complete freedom from need for tricuspid valve reoperation. Various technical modifications for valve repair are being tested, such as pericardial septal leaflets, cone repairs, and leaflet augmentation, in addition to conventional annular plication and sliding leaflet plasties (6,7,10). There are many variants in an Ebsteinoid tricuspid valve, making it important to have more than one repair strategy in a surgical armamentarium. As a society, our expertise in this area will continue to progress.

**Conclusions**

Long-term outcomes for patients with Ebstein anomaly demonstrate that functional status problems are related not only to the tricuspid valve but also to the fundamental problem of Ebstein’s myopathy. The time appears to have arrived for systematic testing of the adjunctive surgical strategies of right ventricular volume reduction surgery, cavo-pulmonary connection, and prophylactic atrial arrhythmia surgery. They hold the potential for reducing late problems of atrial arrhythmia and right ventricular myopathy. Continued improvement in valve repair strategies will provide the opportunity for decreased need of reoperation on the tricuspid valve.
REFERENCES


