

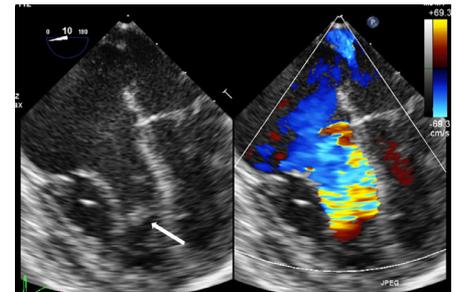
Two Sides of a Coin



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The atrioventricular (AV) valve is both anatomically and developmentally related to the underlying ventricle. Congenital developmental defects of AV valves, therefore, frequently impact the underlying ventricle as well, and vice versa. Ebstein's anomaly is thought to result from defective delamination of the tricuspid valve (TV) resulting in apical displacement of the valve leaflet. The asymmetric leaflet orientation results in poor leaflet coaptation and tricuspid regurgitation (TR, Fig. 1). Several lines of evidence suggest that the underlying right ventricle (RV) is not normal in these patients. In the most severe form of the disease, the RV is unable to maintain antegrade pulmonary blood flow, such that, even in the presence of an anatomically well-developed pulmonary valve, neonates usually present with functional pulmonary atresia. These hearts are incapable of supporting bi-ventricular circulation, and TV exclusion with single-ventricle palliation is necessary for long-term survival.^{1,2} Effective RV remodeling requires decompression of the dysfunctional RV into the low-pressure right atrium.²

Patients with less severe forms of the disease present later in life and symptoms are primarily related to the severity of TR. A plethora of techniques has been described to repair the abnormal TV in Ebstein's anomaly. The cone repair³ has rapidly gained popularity as a durable and reproducible method to repair the abnormal TV. The goals of repair include complete delimitation of the leaflets, repositioning the leaflets in the true tricuspid annulus, and ensuring adequate leaflet coaptation to ensure valve competence. The "atrialized" portion of the RV, which is located on the ventricular side of the now repositioned TV, is a poorly functioning region of the RV. It adds little to the contractile effort, but more importantly, can act as a dyskinetic capacitance chamber impeding effective antegrade blood flow. Hence, plication of this atrialized segment is undertaken at the time of TV repair. Traditionally, this has been accomplished from within the right atrium across the TV. This approach provides excellent visualization of the RV architecture and sutures



Preoperative transesophageal echocardiogram of a child with Ebstein's anomaly.

Central Message

As surgical results of tricuspid valve repair for Ebstein's anomaly continue to improve, long-term outcomes rely on effective ventricular remodeling and function.

can be placed with full exposure to the repositioned TV leaflets. However, the intracardiac approach puts coronary arteries on the free wall of the RV on the blind side and, hence, at risk for suture injury. Careful plication with meticulous attention to the epicardial surface to prevent entrapment of coronary vessels is crucial. In this issue of the journal, Srivastava et al⁴ report an external plication technique to address the atrialized RV during TV repair for Ebstein's anomaly. The authors externally visualize an inverted triangular-shaped atrialized RV prior to initiating cardiopulmonary bypass. Subsequently, on bypass with the heart empty and beating, they perform transmural plication and then proceed with cardiac arrest and TV repair. As the authors point out, this technique allows continued visualization of perfused coronaries during plication, even though in 1 of their 2 patients, they had to revise the repair due to coronary compromise. In contrast, it puts the TV leaflet on the blind side of the plication. Although the authors claim that staying 1–2 mm within the apex of the so-called inverted triangle will avoid entrapment of usable TV tissue, this may not universally be the case. Not infrequently, there is nondelaminated valve tissue above the false annulus that we have been able to delaminate and incorporate into our repair. Also, long-term follow-up is required to confirm that their method leads to reliable RV remodeling and to verify the authors' speculation that transmural plication somehow electrically isolates the atrialized RV and would thereby result in reduced arrhythmia. Regardless of these shortcomings, the

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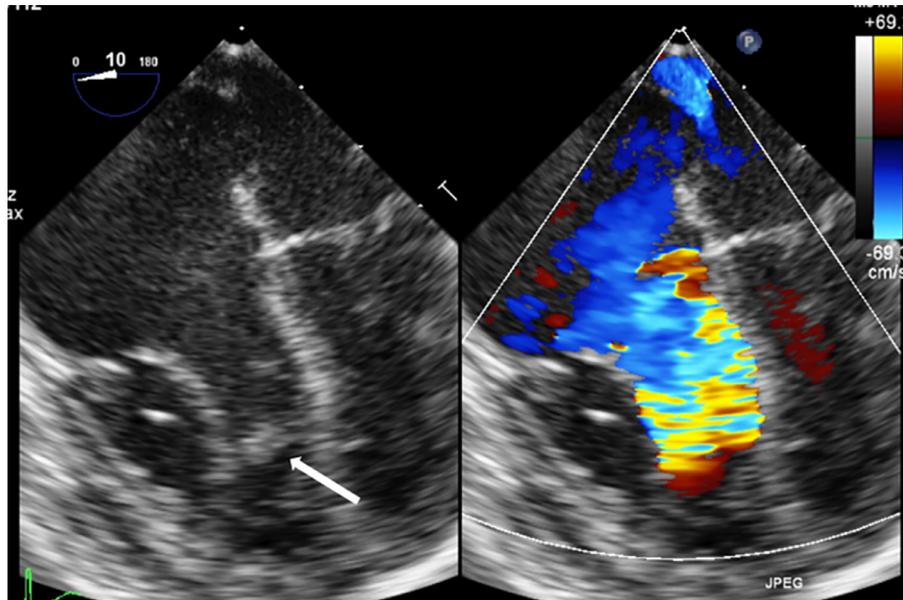


Figure 1. Preoperative transesophageal echocardiogram of a child with Ebstein’s anomaly and apical displacement of tricuspid valve (arrow in left panel) with severe tricuspid regurgitation (right panel).

authors should be congratulated for an elegant and novel approach that looks at RV plication from a very different angle, quite literally.

As surgical results of Ebstein’s repair continue to improve, attention should turn to the long-term functional outcomes of the RV. Several excellent studies have evaluated physiological outcomes following cone repair.^{5,6} There is reliable reduction in grade of TR, reduction in right atrial dimension, and improvement in antegrade pulmonary blood flow. Yet, RV function does not uniformly improve, and, despite symptomatic improvement, exercise tolerance also does not predictably recover in all patients.⁶ These sobering findings reiterate that Ebstein’s anomaly not only affects TV leaflets, but also variably impacts intrinsic RV properties. Is there a way to predict apriori which RV is likely to struggle in the long term? Is there a molecular variable that can be used to determine prognosis? Are there novel approaches to augment RV function for the long term? Research to answer these questions is already well underway. A worthy example is the use of mononuclear cells isolated from the bone marrow. Following successful preclinical evaluation, the effect of intramyocardial injection of these cells into the RV at the time of Ebstein’s repair is being tested

in a clinical trial.⁷ In this era of effective surgical techniques, successful long-term outcome in children with Ebstein’s anomaly relies on our ability to ultimately impact RV remodeling and function.

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