

complete anatomic RV that consists of both aRV and fRV preoperatively.³

5. The LV end-diastolic volume increased postoperatively owing to the ventricular septal shift back toward the RV after surgery and better forward flow from the RV, resulting in improved LV preload. The LV end-systolic volume did not show a significant change.

The major limitation of this study is the small number of patients that had follow-up CMR imaging. Although it is well recognized that CMR imaging can provide valuable data and is superior in quantifying ventricular volumes to echocardiography, the routine use of CMR scans in patients who are doing clinically well might not be justifiable given the cost, the potential need for general anesthesia in small children, and the fact that most surgeons rely on echocardiography to guide feasibility and strategies for tricuspid repair. This highlights the need for a prospective CMR imaging study for patients with EA undergoing cone repair,

preferably as a multicenter study to provide a definitive assessment of late outcome regarding RV remodeling. Assessment of RV deformation by feature tracking on CMR imaging also may provide insight into the RV myopathy of EA. Such natural history data are imperative for assessing adjunctive therapeutic options for RV myopathy, for example, cell-based therapy.⁴

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Commentary: Cone reconstruction for Ebstein’s anomaly is here to stay

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Cone reconstruction of the tricuspid valve to treat valvular regurgitation in Ebstein’s anomaly has now become the accepted method for reproducibly improving tricuspid valve function. Since the early description of the cone technique and initial results reported by DaSilva and colleagues,¹ several centers have adopted the technique and

CENTRAL MESSAGE

Cone reconstruction of a regurgitant Ebstein’s tricuspid valve is associated with improved valve function and global biventricular function and filling late after surgery.

reported their early and mid-term results.²⁻⁴ The basic concept of the cone procedure relies on mobilization and clockwise rotation of valve tissue to create a cone-like valve morphology with valve tissue attachment at the anatomic atrioventricular junction and support cords originating from the right ventricular apex. Reduction or plication of

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the anatomic annulus along with the atrialized portion of the right ventricle is also performed to reduce the valve orifice size closer to normal. While the main goal of the operation is to reduce valvular regurgitation, a reduction of right ventricular volume is proposed as a potential long-term benefit.

Early reports on the results of this new technique have focused primarily on valve function and changes in right ventricular volume. The main endpoints reported in these studies have been either echocardiographic assessment of tricuspid valve function or, more recently, cardiac magnetic resonance measures of ventricular volumes and tricuspid valve regurgitant fraction, along with survival.

Although valve function and anatomic or volumetric features are important for determining the impact of valve reconstruction on the right ventricle, there is little evidence that these parameters are correlated with functional capacity. Objective functional metrics with cardiopulmonary exercise testing, such as peak VO_2 and VE/VCO_2 (ventilatory response to carbon dioxide production at the anaerobic threshold), are standard methods for assessing functional capacity, with the former most impacted in patients with Ebstein's anomaly and tricuspid valve regurgitation. However, in children, accurate assessment of peak VO_2 is often not possible owing to the need for patient cooperation. On the other hand, the presence of symptoms is often absent unless there are associated defects, such as atrial septal defects resulting in cyanosis.

Taken together, the inability to identify symptoms related to Ebstein's anomaly of the tricuspid valve, even in the presence of significant valve regurgitation, and the limited ability to obtain objective measures of functional capacity, have resulted in the absence of uniformly agreed-upon indications for surgical intervention. This is particularly true in children. For this reason, many investigators have sought alternative methods of assessing functional capacity to evaluate the impact of the cone procedure on overall performance late after surgery and have attempted to correlate these with objective changes in exercise performance.

Previously, Ibrahim and colleagues⁵ demonstrated improved forward pulmonary blood flow per heartbeat (effective right ventricular stroke volume) and increased left ventricular diastolic filling indexed to body surface area after cone reconstruction, and this was associated with improved peak oxygen uptake on a bicycle ergometer in a subset of young adults with near-term follow up (2.7 years). In this more recent update from the same group at Great Ormond Street Hospital for Children,⁶ they have

expanded their data set and duration of follow-up, now evaluating global ventricular function late after cone reconstruction. The most significant findings are that both right ventricular and left ventricular effective stroke volume are increased in these patients at a median follow-up of 5.1 years, with the latter parameter more closely associated with exercise functional capacity. The significance of this finding is twofold: (1) the cone reconstruction for regurgitation in Ebstein's anomaly of the tricuspid valve results in long-term improvement in global ventricular function, and (2), magnetic resonance imaging (MRI) measurements of ventricular performance are likely to be better indicators of exercise capacity than anatomic measurements, such as end-diastolic volume index.

These findings support the notion that another indication for surgical reconstruction of the regurgitant Ebstein's tricuspid valve is to improve left ventricular filling and thereby, increase effective left ventricular stroke volume. Thus, when the gold standard of functional exercise capacity is not available to assess the impact of tricuspid regurgitation in Ebstein's anomaly because of patient age, MRI measurement of not only tricuspid regurgitant volume but also, and equally important, of left ventricular performance is an excellent surrogate. With growing evidence that repairing longstanding tricuspid valve regurgitation in association with poor biventricular function does not result in recovery of global ventricular function, indices to determine objective indications for early intervention are likely to mitigate this problem.

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