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Editorial

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Cone Reconstruction for Ebstein's Anomaly: Is It Durable in Pediatric Patients?

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▸ See the article "Long-Term Outcomes of Modified Cone Reconstruction for Ebstein's Anomaly in Pediatric Patients in a Single Center" in volume 54 on page 78.

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Various surgical repair techniques have been introduced to repair Ebstein's anomaly. In most of the procedures, their long-term surgical outcomes were unsatisfactory with a high rate of recurrent tricuspid regurgitation (TR) and reoperation.[1] Recently, cone reconstruction has been widely accepted as a preferred procedure to repair Ebstein's anomaly. By constructing a cone-shaped valve with full leaflet-to-leaflet coaptation, cone reconstruction can achieve nearly anatomic restorations of the tricuspid valve anatomy and function. Compared to various other procedures, favorable outcomes of cone reconstruction have been reported in adult patients.[2] Cone reconstructions are also applied for children and even for neonates and the outcomes are favorable.[3][4] However, tricuspid reoperation is more frequent in young children.[5] Therefore, there are still questions to be answered to adopt this technique in small children. Is cone reconstruction durable in small children? Does the reconstructed tricuspid valve grow accordingly as the child grows? Does cone reconstruction at a younger age help to reverse the remodeling of the ventricles?

In this issue of the Journal, Park et al.[6] reported a result of cone reconstruction for Ebstein's anomaly in pediatric populations. It is a retrospective review of single center, single surgeon experience. Fourteen pediatric patients (age <18 years) were included. The median age at the operation was 5.8 years (range, 1.01–16.6) and 43% of the patients were younger than 4 years old. Seventy-eight percent of the patients had Carpentier's type C or D. There was no early or late mortality. Follow-up duration was 7.6 years (interquartile range, 4.9–11.3). Key findings are followings: (1) Tricuspid valve function could be effectively restored in pediatric patients and remained good for up to 10 years. Only one patient (7%) had reoperation at postoperative 6 months for leaflet perforation. None of the other patients had more than moderate TR through the follow-up. (2) The tricuspid valve grew appropriately according to the patient's growth. No patient had more than mild tricuspid stenosis during follow-up. Serial echocardiography demonstrated that the z value of the tricuspid valve annulus remained between –1 and +1 for 7 years. (3) Cone reconstruction was a durable procedure even in small children. As described before, the tricuspid valve function remained good and the right atrial and ventricular size remained unchanged during follow-up. Body surface area indexed right atrial end-systolic area and right ventricular (RV) end-diastolic area measured by echocardiography decreased after the operation, and it remained unchanged. (4) Left ventricular (LV) reverse remodeling occurs over time after cone reconstruction. LV end-systolic, end-diastolic volume, and stroke volume increased postoperatively, and it increased progressively for up to 3 years.

Data Sharing Statement

The data generated in this study is available from the corresponding author upon reasonable request.

The major limitation of this study is that cardiac chamber dimension and function were assessed by echocardiography. Currently, cardiac magnetic resonance imaging (MRI) is considered the gold standard for evaluating cardiac function and dimensions. With no MRI assessment, the authors could not present data regarding the RV function and volumetry. Recently Neijenhuis et al.[7] reported changes in ventricular function and cardiac chamber size after cone reconstruction using cardiac imaging (MRI). In their data, TR fraction decreased, RV ejection fraction remained stable, antegrade pulmonary arterial beat volume, LV stroke volume, and antegrade aortic beat volume increased. The LV stroke volume improved progressively with time since surgery. Right atrial size decreased and functional RV and left heart area increased. RV volumes showed a tendency to normalize. However, in their study, patients were older and the follow-up duration was not long enough to answer the questions regarding the effect of cone reconstruction and its durability in small children. In clinical practice, there are huddles to take MRI in young children, especially when the patient is asymptomatic. However, further studies with a large number of patients assessed with cardiac MRI are necessary to study biventricular reverse remodeling. Moreover, the effect of restored tricuspid valve and biventricular function on the cardiopulmonary exercise capacity and long-term clinical outcome should be investigated. These studies will be the base to determine the indication and optimal timing of surgical repair in young, especially asymptomatic children with Ebstein anomaly.

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