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Commentary: Staged cone repair for Ebstein anomaly

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CENTRAL MESSAGE

Patients with Ebstein anomaly who received a tricuspid valve patch closure at birth may be a candidate for a staged tricuspid valve repair leading to a 1.5- or 2-ventricle circulation.

In this edition of *JTCVS Techniques*, Da Silva and colleagues describe a "cone" repair of the tricuspid valve in 2 patients with severe Ebstein anomaly (EA) who had previously undergone right ventricle (RV) exclusion at birth.[1] Rather than following a single-ventricle palliation pathway, 1 patient was converted to a one-and-a-half ventricle circulation and the other was converted to a 2-ventricle circulation as result of the described techniques. The authors postulate that cone repair following RV exclusion may effectively balance early survival in this challenging lesion, avoiding the long-term risks of single-ventricle palliation.

Severe EA in the neonate remains a challenging lesion despite improvement in operative techniques. Early surgical intervention in the neonate with severe EA consists of biventricular repair or single-ventricle palliation. Biventricular repair consists of tricuspid valve repair, reduction of the right atrium, establishment of unobstructed egress from the RV to the pulmonary artery (PA), and partial closure of the atrial septal defect.[2] In contrast, the first stage of single-ventricle palliation consists of fenestrated patch closure of the tricuspid valve, division of the pulmonary artery, atrial septectomy, and placement of a systemic to PA shunt.[3] The decision to pursue single or biventricular repair in the neonate rests on the adequacy of the RV, amount of tricuspid valve tissue available, and presence of functional or anatomic pulmonary atresia. Single-ventricle palliation has been described with good early survival and encouraging late survival following second- and third-stage palliation[3]; however, long-term risks of single-ventricle physiology remain unavoidable. Primary 2-ventricle repair in the neonatal or early infancy period has had variable results, with particularly high rates of early mortality in patients with severe EA and pulmonary atresia.[4]

In the report by Da Silva and colleagues, following neonatal RV exclusion, 1 patient underwent bidirectional Glenn and pulmonary valvotomy at 4 months, before tricuspid valve "cone" repair at 17 months. The other patient was not felt to be a good candidate for bidirectional Glenn due to elevated mean PA pressures, elevated left ventricular end-diastolic pressure (15 mm Hg), pulmonary vein stenosis, and mechanical ventilator dependence. As such, this patient underwent tricuspid valve "cone" repair, pulmonary valvotomy, and repair of pulmonary vein stenosis at 5 months. Both patients survived to discharge following "cone" repair with excellent hemodynamics, no oxygen requirement, no evidence of cyanosis, and echocardiographic results that demonstrated competence of the repaired tricuspid valve with adequate RV function.

Da Silva and colleagues present a novel addition to the management of the complex neonate with severe EA. Primary RV exclusion, followed by reassessment of the RV, pulmonary valve, and left ventricle, and consideration for a staged "cone" repair,[5] in which the leading edge of the mobilized anterior and posterior leaflets are rotated

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clockwise and sutured to the septal surface of the anterior leaflet to form a cone within the RV, provided excellent tricuspid valve competence on postoperative echocardiogram. The early results of Da Silva and colleagues suggest that a patient need not necessarily undergo continued single-ventricle palliation after undergoing an RV exclusion procedure as a neonate. The RV may recover function and become adequate through early infancy. Long-term follow-up regarding the qualitative function of the RV will be critical, as will further elucidation on which patients should undergo "cone" repair following RV exclusion.

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