Editorial

Neonatal Ebstein repair—where are we now?

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Ebstein anomaly is a cardiomyopathy characterized by arrested delamination of the tricuspid valve leaflets. This results in displacement of the hinge points of the leaflets into the right ventricle (RV) cavity. The right ventricular myocardium, and in 10–15% the left ventricular myocardium, will have varying degrees of abnormalities in structure and function. The wide anatomic spectrum of Ebstein anomaly results in a highly variable clinical presentation ranging from the severely symptomatic newborn to the asymptomatic adult. Most infants with Ebstein anomaly and mild or moderate cardiomegaly will be asymptomatic at birth. However, when severe cardiomegaly, severe tricuspid regurgitation, and additional cardiac lesions coexist, the neonate can present with profound cyanosis, severe heart failure, and shock. Further complicating the management of the symptomatic neonate are elevated pulmonary vascular resistance, compromised lung volumes due to severe cardiomegaly and reduced effective RV size and function. This combination of findings can result in functional pulmonary atresia if the RV is inadequate to provide antegrade pulmonary blood flow. Despite maximal medical therapy using prostaglandin infusion to maintain ductal patency, inhaled nitric oxide to lower pulmonary vascular resistance, and inotropic support, some patients will require surgical intervention in the neonatal period. Surgical outcomes for neonatal Ebstein repair continue to be challenging although recent reports are demonstrating improved survival. Neonatal surgical options include tricuspid valvuloplasty, reduction atrioplasty and subtotal closure of the atrial septal defect (biventricular repair), or single ventricle palliation ultimately resulting in a Fontan procedure.

Kumar *et al*. recently reported long-term outcomes for a single ventricle approach utilizing the modified Starnes procedure for neonatal Ebstein palliation (1). This approach consists of right ventricular exclusion with a fenestrated patch closure of the tricuspid valve, right atrial reduction and placement of a systemic-to-pulmonary artery shunt. In addition, MPA ligation is performed if significant pulmonary regurgitation is present. They reported long-term outcomes for 27 neonates; 22 survived to hospital discharge. No fenestration was placed in the tricuspid valve patch in the initial three patients and two of those died. Subsequently, 24 patients had a fenestrated tricuspid valve patch to allow for decompression of the RV. There were three early deaths in this group. For those with fenestrated RV exclusion the 1-, 5-, and 10-year survival was 87%, 87%, and 81%, respectively. Of the 22 surviving neonates, 20 underwent Fontan completion with a 1-, 5-, and 10-year survival of 95%, 95%, and 89%, respectively. These outcomes are impressive since the majority (22 of 27 patients) had anatomic or functional pulmonary atresia and an average Great Ormond Street score of 1.4 (1.3–1.8), indicating a high risk subgroup. The advantages of the modified Starnes procedure include: exclusion and decompression of the RV, remodeling of the RV within 2 weeks resulting in a smaller RV and preservation of normal left ventricular function. Kumar *et al*. established that fenestration of the exclusion patch is necessary, but RV plication is not needed (1). RV plication adds time to the procedure without impacting RV

remodeling. Long-term outcomes for the modified Starnes procedure in neonates with severe Ebstein anomaly are similar to outcomes for other complex lesions that require neonatal intervention.

By comparison, Knott-Craig and Goldberg showed improved survival with biventricular techniques applied during the neonatal period. The operation they described included tricuspid valvuloplasty, reduction atrioplasty and subtotal closure of the atrial septal defect (2). In 2011, Boston *et al*. updated their ongoing experience with biventricular repair in 32 young patients, 22 were neonates and 21 had neonatal biventricular repair with a mean follow-up of 5.9 years. The longest follow-up was 16 years. Early survival among the neonates was 69.5%, with one late death. Of note, early survival for neonates with anatomic pulmonary atresia was only 40%, compared to 90% in those neonates with more normal pulmonary valves (3).

In the current era, surgical intervention is most often advised in early childhood or adolescence and the operation focuses on tricuspid valve repair. Numerous valve repair techniques have been described and range from monocusp techniques to circumferential leaflet reconstruction—the cone repair. The cone repair involves surgical delamination and mobilization of tricuspid valve leaflets in order to reestablish leaflet-to-leaflet coaptation and re-anchoring of the reconstruction at the level of the atrioventricular groove. In addition, the atrialized RV is plicated with right reduction atrioplasty and closure or subtotal closure of the atrial level shunt. Bidirectional cavopulmonary shunts and arrhythmia surgery are applied selectively (4,5). In the initial Mayo Clinic experience of 84 patients younger than 21 years of age, 98% had a successful tricuspid valve repair by hospital discharge with only one death (a neonate with severe pulmonary disease) and there was only one valve replacement (6). The cone repair requires extensive and complex leaflet manipulation and is relatively contraindicated in the presence of moderate pulmonary hypertension. Thus, the cone repair should be applied with caution in the setting of a critically ill neonate with preference toward a simpler and expeditious monocusp repair.

The goal in neonates with severe Ebstein anomaly should be initial medical management, including measures to reduce the pulmonary vascular resistance and to maintain adequate pulmonary blood flow with prostaglandin infusion. Although management strategies should be tailored to surgeon and institution experience, general guidelines include the following: if the RV dysfunction prohibits weaning from PGE infusion then the modified Starnes procedure appears to be a favorable approach, particularly if anatomic/functional pulmonary atresia is present. Biventricular repair is selected when tricuspid valve anatomy is favorable for repair and the clinical situation is more stable. The importance of reduction atrioplasty cannot be overemphasized to allow adequate room for lung expansion and ventilation. Late survival is excellent for patients treated with either surgical pathway if they survive the neonatal operation.

This is an exciting era for those who care for patients with Ebstein anomaly. There have been great advances in the neonatal intensive care management and improving surgical techniques available to address this lesion. The cone reconstruction allows for the most anatomic repair and may be the operation of choice beyond the neonatal period; early results are excellent and late results are encouraging. The need for cardiopulmonary bypass in the neonatal period optimally should be avoided. But, when it is necessary, the modified Starnes procedure can provide excellent results and may be preferred to a biventricular repair for the most difficult patients with Ebstein anomaly, or when there is limited experience with tricuspid valvuloplasty techniques in the newborn.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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