Commentary: Neonatal Ebstein: Starnes Procedure First…Fontan Not Necessarily the Last

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CENTRAL PICTURE Legend: The authors Elizabeth H. Stephens, MD, PhD (R) and Joseph A. Dearani, MD (L).

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The Starnes procedure can be instrumental in stabilizing critically ill neonates with Ebstein’s anomaly, but Fontan palliation does not always need to be the final step.

Cleveland and colleagues present a case of a critically ill neonate with Ebstein anomaly with an accompanying thoughtful discussion. The key strengths of the paper include successful management of this premature, IUGR baby through the neonatal period (although the patient remains hospitalized at time of paper submission) and the center’s focused approach and successful track record on the Starnes procedure for management of neonatal Ebstein anomaly. However, their unique strategy for a long central shunt many surgeons may find controversial, particularly in a low-birth-weight baby, where the ideal length is not certain, and optimization of pulmonary/systemic blood flow would be difficult.

Ebstein anomaly, when diagnosed in fetal life, carries a significant risk of perinatal mortality. Recent findings from a multicenter study indicate that the likelihood of fetal demise or neonatal death is approximately 45%. Analysis revealed that certain hemodynamic risk factors play a crucial role, namely the absence of forward blood flow through the pulmonary valve or the presence of pulmonary regurgitation at the time of diagnosis. Notably, the presence of pulmonary regurgitation accompanied by a circular shunt is the most influential predictive factor for perinatal mortality.

A comprehensive investigation conducted by Holst and colleagues, using the Society of Thoracic Surgeons database, explored the surgical treatment of neonatal Ebstein anomaly. The study revealed an overall mortality rate of 27%, with tricuspid valve repair specifically contributing to a mortality rate of 40%. These poor results underscore the pressing need for ongoing improvements in managing this challenging patient population. The high mortality rates can be attributed in part to the presence of a circular shunt resulting from both pulmonary and
tricuspid regurgitation. The need for early surgical intervention in this setting is inevitable and usually occurs in the setting of metabolic acidosis, severe hypoxia, and low cardiac output syndrome with hemodynamic instability.

The Starnes procedure serves to alleviate the burden on the right ventricle by decompressing it, pulmonary regurgitation is eliminated with ligation of the pulmonary artery, fenestrated patch closure of the tricuspid valve ensures optimal decompression of the right ventricle while maintaining the coronary sinus in the RA, and reduction atrioplasty facilitates lung expansion and development. These maneuvers enhance the left ventricle's capacity to adequately supply both systemic and pulmonary circulations. But most importantly, the Starnes procedure represents the most predictable and consistent operation for pediatric cardiac surgeons who see only 1-2 newborns with Ebstein each year thereby limiting their ability to gain experience with a biventricular repair with complex tricuspid valvuloplasty.

The Starnes approach has historically led patients down the path of single ventricle repair, eventually culminating in the Fontan procedure. However, it is now possible to revisit patch tricuspid closure later in infancy and pursue tricuspid repair, employing techniques such as monocusp repair or, preferably, the cone technique to rehabilitate the right ventricle (RV) and achieve a 1.5 or 2-ventricle repair. When considering the cone repair after the Starnes procedure, it is advisable to utilize a Gore-Tex patch to exclude the RV, as it is associated with fewer adhesions to the leaflets compared to bovine pericardium. This choice facilitates the later takedown of the patch during the cone repair. Additionally, the patch should be sutured above the tricuspid valve (TV) annulus and along Todaro's ligament along the septal area, while ensuring that the coronary sinus remains draining into the right atrium. These technical measures are aimed at enabling the removal of the patch without causing damage to the TV leaflets or the atrioventricular node when transitioning to a 1.5 or 2-ventricle circulation.

As enthusiasm to avoid a single ventricle pathway increase, issues to be considered include the following: the RV involutes with fenestrated tricuspid valve closure and can get smaller in a short timeline. Keeping RV size in mind, the optimal timing to conversion needs to be carefully monitored so intervention is performed before the RV becomes too small, particularly if a 2-ventricle circulation is the goal. The second issue is technical – what is the best way to handle pulmonary artery ligation when pulmonary atresia is functional (i.e., not anatomic pulmonary atresia). Ideally, it would be preferable to preserve the native pulmonary valve and avoid a RV-PA conduit at the time of conversion. Additional experience is necessary to provide clarity on these issues.

When contemplating the optimal surgical management for critically ill neonates with EA, it is worth noting that the Starnes operation can be replicated in most pediatric heart centers and consistently provides a desirable physiological outcome. Many are now regarding this procedure as the preferred choice for unstable newborns with Ebstein anomaly. On the other hand, cone
repair later in infancy or following the Starnes procedure can be accomplished in experienced centers. Furthermore, it is important to recognize that Starnes palliation no longer necessitates a commitment to a Fontan circulation. Instead, the Starnes procedure serves to ensure the survival of critically ill neonates, preparing them for a more definitive and anatomic procedure with a separated circulation in the future, when their clinical condition is more stable. The Fontan procedure is not necessarily the final pathway after Starnes palliation.

References;
