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Commentary: The best of both worlds? Conversion of a neonatal modified Starnes procedure to biventricular circulation

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CENTRAL MESSAGE

Performance of a cone procedure after a neonatal modified Starnes may optimize long-term outcomes by providing effective early palliation with a long-term biventricular circulation.

The Starnes procedure has enabled the salvage of neonates with severe Ebstein anomaly, even those presenting in cardiogenic shock.[1] However, it may carry a significant long-term downside by committing a patient to a long-term single-ventricle circulation due to an inability to tolerate the neonatal period of high pulmonary vascular resistance. Although outcomes with Fontan palliation in these patients, especially those with a modified Starnes procedure (fenestrated right ventricular exclusion) are good, there continues to be late mortality that may, in part, be attributable to the myriad negative consequences of the total cavopulmonary connection.[2] The opportunity to use the Starnes procedure to palliate a neonate through the high-risk period while preserving subsequent conversion to a 2-ventricle (or 1½-ventricle) circulation has the potential to improve long-term outcomes in these patients.

In the current issue of *JTCVS Techniques*, da Silva and colleagues[3] describe just such a pathway in 2 patients. There has been increasing interest in using the cone procedure for primary repair of the tricuspid valve in neonates.[4-6] However, some patients may present in extremis and are unlikely to tolerate the longer procedure aimed at repairing the valve in the early neonatal period. In such patients, use of the Starnes procedure may be required to improve left ventricular function and systemic cardiac output while maintaining systemic oxygenation levels. The technique described by the authors of subsequently performing a take-down of the right ventricular exclusion patch and performing a cone procedure suggests that these patients are not inevitably headed down a single-ventricle palliation pathway forever.

The flexibility of the approach by da Silva and colleagues is perhaps its greatest advantage. One of the primary challenges with Ebstein anomaly is the clinical heterogeneity and the challenge of selecting the appropriate procedure for the appropriate patient, thereby maximizing both short- and long-term survival. The option to restore a biventricular circulation after the cone procedure suggests that decisions made in the first weeks of life need not be irrevocable.

However, this technique may simply be pushing challenging decisions into later childhood. A case series of 2 patients cannot begin to answer all of the important questions: who should get this procedure? What preoperative assessments can inform whether a patient will tolerate it? Are there techniques for right ventricular exclusion that will optimize the function of the right ventricle and make sure a repair more likely? Which patients will benefit from conversion to a 1½-ventricle repair using a cone procedure and a superior cavopulmonary connection? These are important questions, and it will take time to accumulate experience.

Despite the unanswered questions, the technique holds promise. By enabling the conversion from the single-ventricle palliation of a Starnes procedure to a long-term biventricular (or 1½ ventricle) circulation, we may be able to achieve the best of both worlds: effective and safe early

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treatment of the neonate in cardiogenic shock with eventual biventricular conversion and avoidance of long-term cavopulmonary palliation.

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