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Commentary: Hypoplastic left heart syndrome with intact atrial septum: Planning for success

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CENTRAL MESSAGE

A multidisciplinary planned approach is critical to successfully manage hypoplastic left heart syndrome with an intact atrial septum.

Over the past few decades, major advances in the form of surgical technique, catheter-based interventions, and critical care management have resulted in improved outcomes with hypoplastic left heart syndrome (HLHS) palliation. Despite this progress, the subset of HLHS that presents with a highly restrictive atrial septum (RAS) or intact atrial septum (IAS) continues to present a challenge. Both an IAS and RAS will produce left atrial and pulmonary venous hypertension. This is thought to lead to abnormal lung development, with reduction in lung perfusion characterized by congenital pulmonary lymphangiectasis and muscularization of pulmonary veins. This has been noted to occur as early as 23 weeks of gestation with histologic changes such as hypercapillarization and widened inter-airspace mesenchyme noted at fetal autopsy.[1] Whether these changes are reversible with left atrial decompression, either prenatally by fetal intervention or postnatally, is unknown. Currently, the 1-year survival in the HLHS population with RAS/IAS is low, with survival between 35% and 50% reported in recent case series.[2,3]

In this issue of the *JTCVS Open*, Sood and colleagues[4] present their institutional strategy for the management of HLHS/IAS. Their multidisciplinary approach involves cesarean delivery, airway and line establishment, and transfer to the cardiac catheterization suite within 30 minutes of delivery. Sternotomy and per-atrial balloon septostomy or atrial septal stenting is performed, followed by bilateral pulmonary artery bands and, in the majority of cases, a ductal stent (HS1P). The authors are to be congratulated on organizing a large group of experts and developing a reproducible approach to this critically ill, high-risk group of neonates. The management plan emphasizes, importantly, efficiency with times of 30 minutes from delivery to the catheterization suite and 5 minutes to per-atrial access. The outcomes on 15 neonates with HLHS/IAS briefly presented are notable, with 10 being discharged home after HS1P and 7 undergoing successful Norwood.

Although the authors present a comprehensive plan, there are a few particulars that would be helpful in providing the complete picture. With regard to the atrial septal intervention, details regarding time to adequate septostomy, failure rate, and indication for surgical conversion would be helpful in understanding the limitations of this strategy. In addition, information about the reasons for mortality and failing to progress to the next stage may provide insight into the challenges these neonates may face secondary to pulmonary venous disease or unfavorable anatomy. We suspect that these specifics will be clarified in a future publication reporting on the outcomes of their strategy.

In closing, we are in full agreement with the authors in the need for institutions to have a multidisciplinary strategy to optimize the outcomes of high-risk neonates with HLHS/IAS. Our strategy has been to have elective cesarean delivery within the women's center inside the children's

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Disclosures: The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Received for publication March 13, 2020; revisions received March 13, 2020; accepted for publication March 20, 2020; available ahead of print April 22, 2020.

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JTCVS Open 2020;1:59-60

2666-2736

https://doi.org/10.1016/j.xjon.2020.03.002

hospital. The newborn is resuscitated, prostaglandins are initiated, and the patient is transported immediately to the cardiac operating room, where the surgical team is standing by. Cardiac anesthesia intubates and places venous and arterial lines expeditiously. A surgical atrial septectomy is done followed by bilateral pulmonary banding. We favor tight bilateral pulmonary artery banding (1.8-2.2 mm), using supplemental oxygen if needed, as even mild pulmonary overcirculation is poorly tolerated in these patients.[5,6] We have kept these patients on prostaglandins for ductal patency and delayed the Norwood operation for at least 4 to 6 weeks to achieve a period of lung recovery. This period is geared toward nutrition, lowering respiratory support, and optimizing pulmonary function with diuresis and dual pulmonary hypertensive medications. This strategy has been used in 4 neonates with HLHS/IAS, with all undergoing Norwood followed by bidirectional cavopulmonary shunt (1 mortality after Glenn). How these high-risk babies will fare long term remains to be seen, but developing an institutional plan is undoubtedly the first step toward success.

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