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Commentary: It takes a village to manage a child with hypoplastic left heart syndrome and intact or highly restrictive atrial septum

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CENTRAL MESSAGE

Management of HLHS and intact atrial septum requires an organized team approach that starts with fetal diagnosis and intervention. Research and innovation are needed to improve early and late outcomes.

In the current issue of the *Journal*, Sood and colleagues[1] from Ann Arbor, Michigan, describe their institution's approach to neonates born with hypoplastic left heart syndrome (HLHS) and highly restrictive or intact atrial septum. Their collaborative multidisciplinary strategy begins with prenatal identification of patients at risk and referral of proper candidates for fetal intervention (creation of atrial communication, stenting preferable over balloon dilation), planned cesarean delivery with rapid resuscitation and expeditious transfer (within 30 minutes) to the catheterization laboratory for sternotomy and per-atrial balloon septostomy and/or stenting, followed by branch pulmonary artery banding and ductal stenting. Concomitant lung biopsy is obtained. Norwood operation is performed at 6 to 8 weeks of age with the aim for hospital discharge in the interim if possible. Their experience includes 15 patients. Hospital survival was 10 of 15 (67%), with 5 of 6 (83%) of those who had fetal intervention surviving the initial intervention. Of those 10, 7 survived to Norwood, with 1 dying after Norwood and 6 remaining alive (5 received Fontan).

Highly restrictive or intact atrial septum is present in 6% to 11% of patients with HLHS.[2-5] Management challenges in these patients include immediate instability (severe hypoxemia because of restricted egress of pulmonary venous blood to right atrium and subsequent pulmonary edema) and persistent elevation of pulmonary vascular resistance (pulmonary arterioles and venous thickening, pulmonary lymphangiectasia) affecting both early mortality and later outcomes.[6] Prenatal diagnosis and fetal intervention are therefore valuable not only to improve early stability but also to potentially reverse those damaging developments in the pulmonary vasculature due to left atrial hypertension. Emerging experience from centers seem convincing that fetal intervention should be considered in these patients, as it would likely improve their condition soon after birth until they undergo opening of the septum soon after birth.[3,4,7,8] On the other hand, the effect of fetal intervention on the pulmonary vasculature and later outcomes has not been well studied or demonstrated yet. A number of studies have looked at the effect of fetal intervention on results in patients with HLHS with intact atrial septum and the results varied; while few have shown improved outcomes, others have not.[2-8] The current experience from Michigan suggests better hospital survival in those who had fetal intervention (5/6) but constant attrition later on (only 3/6 alive), and that might suggest better initial stability but persistent issues related to lung development. These findings raise few questions: (1) Should these patients always be referred to centers with fetal intervention capabilities? (2) Should lung biopsy play a bigger role in decision-making in these patients? (3) Is there a role for pulmonary vasodilators?

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While the decision to perform immediate cardiac catheterization to create atrial communication is largely settled, the choice of subsequent intervention seems to vary among institutions. Sood and colleagues elected to perform hybrid stage I (pulmonary artery banding and ductal stenting) and delay Norwood operation for 6 to 8 weeks. The argument to delay Norwood operation is to allow the lungs to recover and have time to assess pulmonary pathology and associated pulmonary vascular resistance. Others have implied different strategies, including Norwood operation few days later, pulmonary artery banding followed by Norwood operation within 1 to 2 weeks, and hybrid stage I followed by a comprehensive second stage months later. Nonetheless, these practices seem to be based on different philosophies and clinical judgment without evidence to support one method over the other. The Michigan results seem comparable with a few other centers that apply different approaches following establishment of atrial communication, all suggesting the need for more research and assessment of optimal strategy. My personal approach is to perform the Norwood operation (using an aortopulmonary shunt) within 2 to 3 days following septal opening and liberal use of pulmonary vasodilators. While this strategy has been helpful in my practice, it is largely driven by personal viewpoint and anecdotal experience.

In short, management of these patients demands considerable joint efforts and collective expertise from multiple specialties, including fetal cardiology, obstetrics, neonatology, interventional cardiology, cardiac surgery, anesthesia, perfusion, and the intensive care unit, along with significant ancillary support systems. Results continue to be suboptimal, suggesting the need for more research and innovation to better understand pathophysiology in these patients and to subsequently revolutionize a successful management strategy that improves both early and late outcomes.

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