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CASE REPORT

ADVANCED

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

Atrial Appendage Anastomosis in Hypoplastic Left Heart Syndrome With Restrictive Atrial Septum

A Novel Approach

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ABSTRACT

Intact atrial septum (IAS), occurring in ~10% of patients with hypoplastic left heart syndrome (HLHS), conveys significant neonatal morbidity and mortality. Perinatal interventions have been described, but outcomes remain poor. We present a fetus with HLHS with IAS who underwent immediate novel postnatal atrial appendage anastomosis, thus achieving rapid left atrial decompression. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:1065–1069)

CASE PRESENTATION

A 31-year-old healthy primigravida was referred for an abnormal 4-chamber view on the fetal anatomy survey. Fetal echocardiography at 20 weeks, 3 days was performed (**Figures 1A to 1D,** Videos 1 to 3).

LEARNING OBJECTIVES

- To understand echocardiographic features that predict progression of fetal AS to HLHS and atrial septal restriction and counseling for expectant parents.
- To identify options for left atrial decompression in HLHS with IAS.
- To recognize atrial appendage anastomosis as a novel approach for left atrial decompression in HLHS with IAS.

QUESTION 1: WHAT OPTIONS EXIST FOR THE PARENTS AND FETUS WHEN A DIAGNOSIS OF SEVERE AORTIC STENOSIS AND EVOLVING HYPOPLASTIC LEFT HEART SYNDROME IS MADE IN MIDGESTATION?

The findings demonstrated severe aortic stenosis (AS) with evolving hypoplastic left heart syndrome (HLHS) and a mildly restrictive interatrial septum. Echocardiographic findings that predict progression of midgestation severe AS to HLHS include reversal of flow in the transverse aortic arch, monophasic mitral inflow, and left-to-right flow across the foramen ovale.[1] Fetal aortic valvuloplasty has been described as a strategy to mitigate progression of fetal AS to HLHS and promote a biventricular outcome.[2] Risks of maternal-fetal interventions include pregnancy loss and preterm delivery. Expectant management with intervention after birth, comfort care at birth, or

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ABBREVIATIONS AND ACRONYMS

AS = aortic stenosis

ECMO = extracorporeal membrane oxygenation

EXIT = ex utero intrapartum treatment

HLHS = hypoplastic left heart syndrome

IAS = intact atrial septum

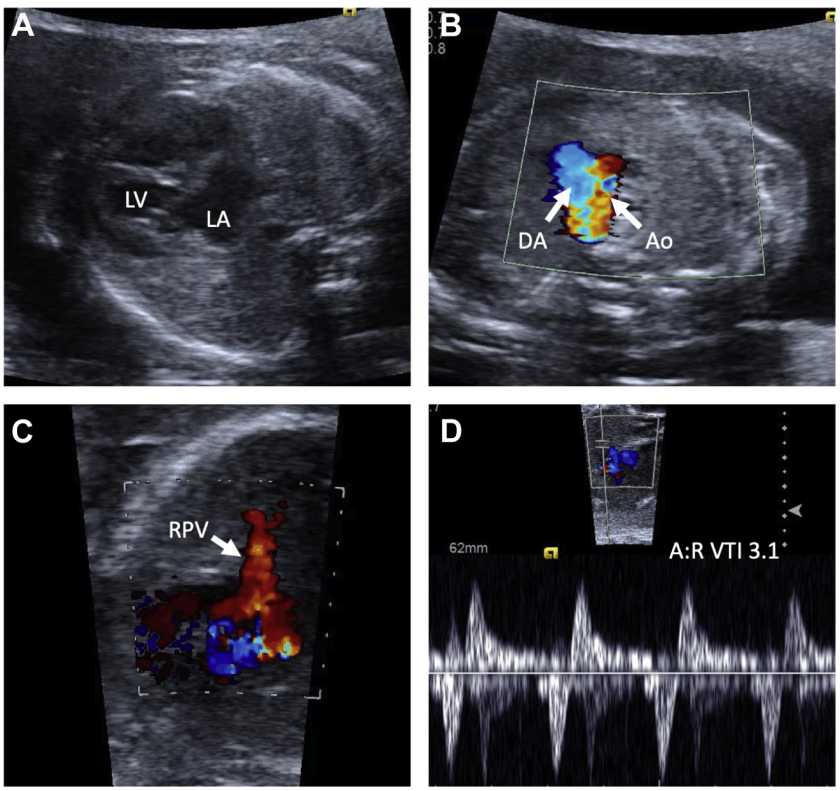
OR = operating room

pregnancy termination were discussed with the couple. A multidisciplinary meeting of obstetricians, neonatologists, surgeons, cardiologists, social workers, and trainees convened, and fetal intervention was discussed as reasonable. The couple were counseled and chose fetal intervention. Fetal aortic balloon valvuloplasty was performed at 21 weeks, 3 days. Postprocedural study showed subjective improvement in forward flow across the aortic valve and no maternal or fetal complications. Subsequent limited evaluations were done at a local hospital, and the patient was re-referred at 34 weeks of gestation given new concern for atrial septal restriction. Follow-up fetal echocardiography was performed at 34 weeks, 2 days (**Figures 2A to 2D,** Videos 4 to 6).

QUESTION 2: WHAT IS THE BEST DELIVERY MANAGEMENT FOR A FETUS WITH HLHS AND INTACT OR SEVERELY RESTRICTIVE INTERATRIAL SEPTUM? WHAT IS THE BEST MANAGEMENT FOR THE MOTHER?

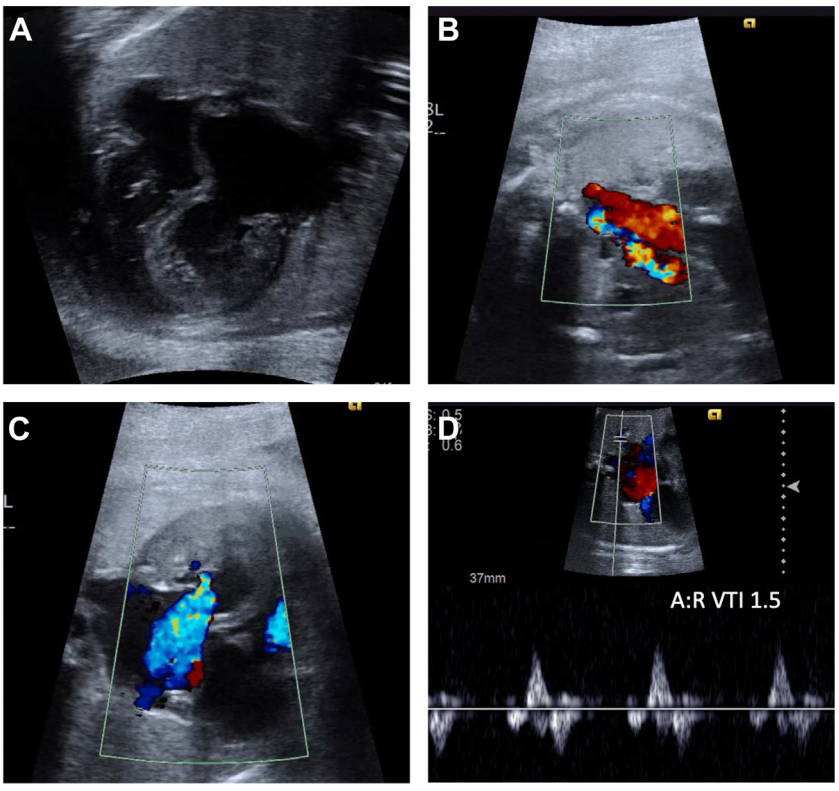
The findings demonstrated ongoing evolution of HLHS and interval development of a severely restrictive atrial septum or intact atrial septum (IAS). Fetal diagnosis of IAS is made by Doppler interrogation of the pulmonary venous flow pattern, with an antegrade-to-reverse velocity time integral ratio of <3 being predictive of need for emergency atrial septostomy.[3] Fetal atrial septoplasty was discussed; however, it was not offered given advanced gestation and the risk for fetal distress necessitating emergency delivery, leaving at best a 34+-week newborn in

FIGURE 1 Fetal Echocardiogram at 20 Weeks, 3 Days



(A) A 4-chamber view demonstrating the atrial septum bowing from left to right and a mildly hypoplastic mitral valve (4 mm; $Z = -2$). The left ventricle (LV) was dilated, with severely diminished function. The aortic valve measured normally (3 mm; $Z = -1.7$), but was thickened, dysplastic, and stenotic (Video 1). **(B)** A 3-vessel view demonstrating retrograde flow in the mildly hypoplastic transverse arch (Video 2). **(C)** and **(D)** Color and pulse-wave Doppler imaging in the right pulmonary vein (RPV) showed an antegrade-to-reverse velocity time integral ratio (A:R VTI) of 3.1:1 (Video 3). Ao = aorta; DA = descending aorta; LA = left atrium.

FIGURE 2 **Fetal Echocardiogram at 34 Weeks, 2 Days**



(A) A 4-chamber view demonstrating a non–apex-forming dilated, hypertrophied left ventricle with significant endocardial fibroelastosis. The mitral valve measured 9.8 mm ($Z = -0.6$). The aortic valve measured 3.1 mm ($Z = -4.8$), peak gradient 90 mm Hg (Video 4). **(B)** A 3-vessel view demonstrating a hypoplastic aortic arch with retrograde flow in the transverse arch (Video 5). **(C)** Color Doppler imaging of the mitral valve demonstrating severe mitral regurgitation (peak gradient 67 mm Hg) (Video 6) and **(D)** pulse-wave Doppler imaging of the left lower pulmonary vein demonstrating and an antegrade-to-reverse velocity time integral ratio (A:R VTI) of 1.5.

extremis with the underlying diagnosis and limited options for resuscitation. A multidisciplinary team consisting of senior representatives from fetal cardiology, cardiothoracic surgery, obstetric and cardiac anesthesia, maternal-fetal medicine, and nursing was convened to review options for coordinating postnatal management. The lowest risk to the mother would be induction of labor at 39 weeks with postnatal cardiac interventional catheterization and operating room (OR) teams on standby during labor and delivery. The lack of control over delivery timing and the potential for urgent cesarean delivery without a ready team and delayed atrial septal decompression were acknowledged. Scheduled cesarean delivery before labor onset with immediate transfer of the newborn to the cardiac OR or catheterization suite would provide the most control. In our center, cesarean deliveries can be performed in a cardiac OR and the infant can be taken to an adjacent OR. Ex utero intrapartum treatment (EXIT) to extracorporeal membrane oxygenation (ECMO) support was not discussed because this option is reserved for fetuses with critical airway compromise in our center, and intrapartum ECMO cannulation was not believed to be superior to expeditious standard surgical cannulation for cardiopulmonary bypass. The parents were counseled about these delivery options and chose the (slightly) higher-risk option to the mother—scheduled cesarean delivery, while being aware of the implications for future fertility and deliveries, as well as the risk for invasive placental disease in subsequent pregnancies. The multidisciplinary team also discussed surgical vs transcatheter approaches, given published success with the latter.[4] Our institutional experience does not favor one approach over the other, but the interventional and surgical teams believed that a surgical approach

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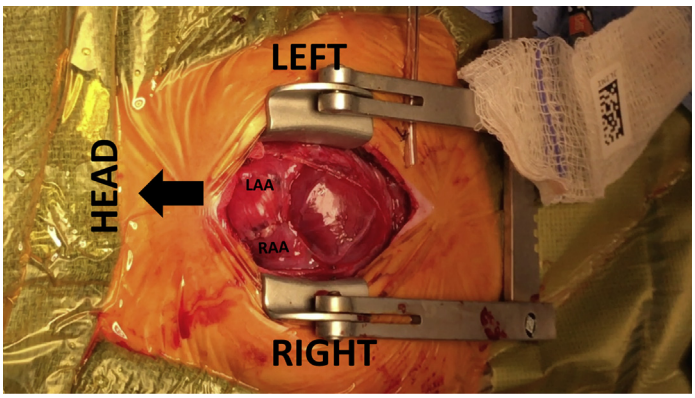
FIGURE 3 Intraoperative Atrial Appendage Anastomosis

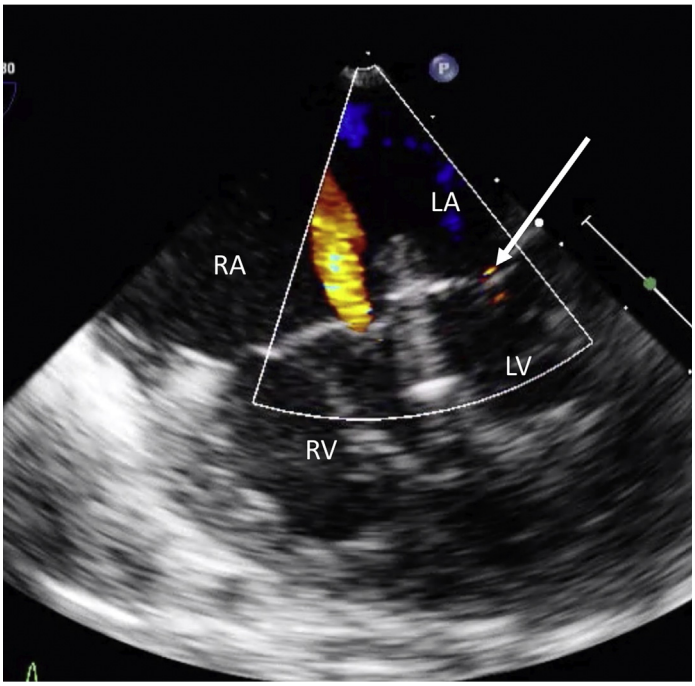


This is also shown in Video 7. RAA = right atrial appendage; LAA = left atrial appendage.

should be planned for because they thought that would be the fastest way to stabilize the infant while also addressing the atrial septum. Options, including echocardiographically guided hybrid stent placement (through sternotomy) and surgical septectomy (using cardiopulmonary bypass), were both planned, with the decision to be made pending assessment after birth.

The fetus was delivered by a coordinated cesarean birth at 37 weeks, 3 days with a planned surgical atrial septectomy. On sternal entry, the right and left atrial appendages were noted to be enlarged and in close proximity. Both appendages were occluded with side-biting clamps and were opened widely. The pectinate muscles were resected, and the appendages were anastomosed together (**Figure 3,** Video 7). On release of the clamps, the patient's oxygen saturations gradually increased from the 50s to the high 80s or low 90s, thus achieving left atrial decompression within 60 minutes of delivery without the use of cardiopulmonary bypass. The neonate was then transferred to the cardiac catheterization laboratory, and transcatheter balloon aortic valvuloplasty was performed to relieve left ventricular obstruction and decrease mitral valve regurgitation. The sternum was left open, and a modified Norwood procedure with a 5-mm right ventricular-to-pulmonary artery shunt was performed 5 days later. Postoperatively, the mitral regurgitation gradually improved without intervention (**Figure 4**). At the time of this report, the patient has a diuretic requirement as a result of pulmonary lymphangiectasia; however, the child is awaiting Fontan completion after recent tricuspid valve repair, right superior cavopulmonary anastomosis, and modified left Blalock-Taussig-Thomas shunt placement.

FIGURE 4 Intraoperative Transesophageal Images During Superior Cavopulmonary Surgery



The mitral regurgitation, which had been severe in utero and was not addressed at the time of the Norwood operation, was now trivial **(arrow).** LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

COMMENT

HLHS is a form of complex congenital heart disease that is managed with single-ventricle palliation, often requiring heart transplantation in later years. Our patient's course was further complicated by in utero development of an IAS. Although HLHS can be generally stabilized at birth with prostaglandins, when a highly restrictive interatrial septum or an IAS is present, the condition can be rapidly fatal after birth because of insufficient egress of oxygenated blood from the left atrium to the systemic circulation. Neonates are at risk for profound hemodynamic instability, acidosis, low cardiac output, and pulmonary edema. Patients with a prenatal diagnosis are recommended to undergo a coordinated delivery with surgical and interventional staff on standby for emergency atrial septectomy or septostomy. Even with carefully coordinated delivery and postnatal intervention, transplant-free survival in HLHS with IAS is significantly worse compared with standard-

risk HLHS, and survivors have significant long-term morbidity.[5,6]

Marshall et al[7] described fetal atrial septoplasty as a strategy for in utero management of HLHS with IAS. These investigators advised intervention at 28 to 30 weeks, a window that allows for the creation of larger defects, yet is early enough to mitigate the development of pulmonary disease.[7] At our institution, atrial stents are offered up to 33 weeks of gestation. Postnatal strategies for management of HLHS with IAS include transcatheter and surgical septostomy or septectomy and ex utero intrapartum treatment (EXIT) procedures. Transcatheter septectomy can be technically challenging because of thick or muscular atrial septa, necessitating transseptal perforation, blade septoplasty, static balloon dilation, or atrial stenting.[4] Surgical atrial septectomy is an option, however requires cardiopulmonary bypass. Because cardiopulmonary bypass is a high risk in HLHS with IAS, EXIT procedures have also been attempted; however limited case reports describe poor outcomes, including neonatal death.[8] Finally, off-bypass atrial septectomy using laparoscopic forceps was described in a small, 3-patient case series, but it has not been widely adopted.[9]

Atrial appendage anastomosis for HLHS with IAS has been described rarely in published reports. We found 1 similar case of left atrial appendage anastomosis to a persistent left superior vena cava in a patient with HLHS with IAS.[10]

CLINICAL PERSPECTIVES

To our knowledge, this is the first published case of atrial appendage anastomosis in a patient with HLHS with IAS. The procedure may not be suitable for all patients. In the absence of juxtaposed atrial appendages, the appendages are anatomically remote. Indeed, the feasibility of the procedure in our patient was likely the result of significant mitral regurgitation causing left atrial appendage dilation. However, this is only a temporizing measure. The anastomosis does not allow for sternal closure, due to risk of obstructing the atrial connection. However, it allows for initial stabilization and some pulmonary recovery and delay of cardiopulmonary bypass with a goal of improvement in overall survival and morbidity.

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KEY WORDS atrial appendage anastomosis, hypoplastic left heart syndrome, intact atrial septum

APPENDIX For supplemental videos, please see the online version of this article.