Innovative two-step management strategy utilizing EXIT procedure for a fetus with Hypoplastic Left Heart Syndrome and Intact Atrial Septum

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**Abstract:**

Hypoplastic left heart syndrome (HLHS) with intact atrial septum (HLHS-IAS) carries a high risk of mortality and affects about 10 percent of all patients with HLHS. Fetal interventions, postnatal transcatheter interventions, and postnatal surgical resection have all been used, but the mortality risk continues to be high in this subgroup of patients. We describe a novel, sequential approach to manage HLHS-IAS and progressive fetal hydrops.

A 28-year-old, G4P2 mother was referred to Mayo Clinic for fetal HLHS. Fetal echocardiography at 28 weeks gestation demonstrated HLHS-IAS with progressive fetal hydrops. The atrial septum was thick and muscular with no interatrial communication. Ultrasound guided fetal atrial septostomy was performed with successful creation of a small atrial communication. However, fetal echocardiogram at 33 weeks of gestation showed recurrence of a pleural effusion and restriction of the atrial septum. The decision was made to proceed with an EXIT (Ex Utero Intrapartum Treatment) delivery and open atrial septectomy. This was performed at 34 weeks gestation under maternal general anesthesia. The fetus was partially delivered and fetal-placental circulation was maintained. A median sternotomy was performed and septum primum was resected. Initial epicardial echocardiogram showed the septal defect continued to be restrictive, so this procedure was repeated and follow up imaging showed a non-restrictive septum. The infant was then intubated, fully delivered, and stabilized before transfer to the intensive care unit. The infant required VA ECMO support on day of life (DOL) 1. The patient later developed hemorrhagic complications leading to discontinuation of ECMO support on DOL 6 and subsequently died on DOL 9 following acute decompensation with bradycardia.

This is the first reported case of an EXIT procedure and open atrial septectomy performed without cardiopulmonary bypass for an open-heart operation in humans and provides a promising alternative strategy for management of HLHS-IAS in select cases.

**Figure 2 composite:**

**A)** Intraoperative picture of surgical team performing open atrial septectomy with the fetus lying on the mother's thighs and attached to mother via umbilical cord. **B)** Postoperative subcostal echocardiographic image showing unrestrictive, surgically created interatrial communication between the left atrium (LA) and the right atrium (RA). SVC = superior vena cava. **C)** Diagram showing the position of the multidisciplinary teams within the operating room during EXIT (Ex Utero Intrapartum Treatment) procedure and atrial septectomy. **1)** Obstetric anesthesiologist. **2)** Maternal fetal surgeon. **3)** Assistant to maternal surgeon. **4)** Pediatric cardiologist. **5)** Cardiac surgeon. **6)** Assistant to cardiac surgeon. **7)** Scrub nurse. **8)** Otorhinolaryngologist.