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The ex utero intrapartum treatment (EXIT) procedure was first used for patients with tracheal compression from CDH.¹ By partially delivering the fetus while it remains attached to the placenta, the EXIT procedure allows time to control resuscitative measures while the infant remains hemodynamically stable. Its use has expanded to include infants with congenital heart disease. We describe an EXIT procedure for a neonate with hypoplastic left heart syndrome with intact atrial septum (HLHS/IAS).

A 36+5 week PGA male with HLHS/IAS was diagnosed prenatally by echo. Prior attempt at prenatal atrial septostomy at an outside hospital failed secondary to impending premature labor. Plan was for an EXIT procedure with intubation and IV access followed by delivery and immediate atrial septostomy. Doppler fetal heart tones were monitored throughout. After uterine incision, the fetus was delivered to the clavicles and intubated. After 2 minutes on placental bypass, severe maternal hemorrhage and fetal bradycardia necessitated delivery prior to IV access. The neonate was transported to the adjacent OR, given endotracheal epinephrine, and underwent sternotomy with catheter-assisted atrial septostomy. Intra-operatively the patient remained stable on low-dose epinephrine infusion; however, pulmonary hypertension post-operatively led to hypotension on days 0 and 1. The patient expired at 13 days from sepsis secondary to NEC.

75 to 84% of patients with HLHS are diagnosed prenatally; 6 to 11% of patients with HLHS also have IAS.² While 27% of cases of HLHS die within 30 days of life, those with HLHS/IAS are universally fatal without fetal or emergent neonatal procedures.³ With an EXIT procedure the time of hemodynamic instability can be maximally reduced, and hopefully increase survival. In our patient, initial atrial septostomy was successful in creating adequate mixing and systemic oxygenation; however, the patient expired due to sepsis from NEC. The occurrence and cause of death is consistent with a recent report stating 47% neonatal mortality for patients requiring emergent post natal intervention for IAS.³

Despite the patient's death, important lessons should be gleaned; preparedness and continuous multidisciplinary communication is essential for a successful EXIT procedure. Anesthesiologists, surgeons, cardiologists, and neonatologists must be aware of the plan and contingencies. Both ORs must be readied ahead of time. Extreme hemodynamic instability is to be expected. All facilities performing EXIT procedures should have an outline of equipment needed for delivery as well as an airway algorithm. In this case, early onset neonatal bradycardia and maternal instability required emergent delivery; however, open communication between team members allowed for a successful surgery.

1. Bouchard et al. The EXIT Procedure: Experience and Outcome in 31 Cases. *Jnl Ped Surg* 37:3. March 2002. 418-426.

2. Vida et al. Hypoplastic Left Heart Syndrome with Intact or Highly Restrictive Atrial Septum: Surgical Experience from a Single Center. *Ann Thorac Surg.* 2007. 84;581-586.

3. Marshall et al. Results of in utero atrial septoplasty in fetuses with hypoplastic left heart syndrome. *Prenat Diagn.* 2008; 28;1023-1028.
