

Title: EXIT to ECMO: A multi-disciplinary tour du force.

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ABSTRACT BODY:

The EXIT (Ex Utero Intrapartum Therapy) procedure is becoming increasingly common in advanced tertiary pediatric care centers for the controlled delivery and intrapartum management of fetuses with life threatening conditions. Extracorporeal membrane oxygenation ECMO is an important part of the treatment algorithm for infants with severe CDH in the United States. It allows for the slow transition from the fetal to neonatal circulation while avoiding the hypoxia associated with pulmonary hypertension. EXIT to ECMO allows for a smooth transition from placental bypass to extracorporeal bypass. We present the management of a fetus with severe congenital diaphragmatic hernia (CDH) that required both the EXIT procedure and immediate ECMO cannulation.

CASE: A 34 year old, G1P0 presented to the fetal center carrying a 24 week gestation fetus with a left CDH and normal chromosomes. The liver and stomach were herniated into the chest and a lung-to-head ratio (LHR) of 1.4 was measured via ultrasound. At the 32 week follow up visit, imaging revealed polyhydramnios with fetal gastric distension and organo-axial gastric volvulus on ultra fast MRI. Poor interval lung growth was noted. The obstructed stomach now had an additional mass effect on the already compressed contralateral lung. There was concern that the gastroesophageal obstruction that accompanies gastric volvulus would not allow gastric decompression and thus further impair ventilation post-partum. This, in the setting of a severe CDH, prompted the consideration for EXIT-to-ECMO.

HOSPITAL COURSE: At 34 weeks gestation, a multidisciplinary meeting was held with representatives from Pediatric Surgery, Obstetrics, Pediatric Anesthesia, Neonatology, Pediatric Cardiology, ECMO technicians, Blood bank and operating room nursing in attendance. The family was introduced to the staff. The indication for the procedure, potential benefits and risks were discussed and the family was walked through the management plan. The roles and responsibilities of each member present were also explained to the family. Surgery was scheduled for 37 weeks gestation. However, two days prior to scheduled surgery she went into labor and the procedure was executed emergently.

INTRAOPERATIVE COURSE: Following aspiration prophylaxis, the patient was positioned supine with left uterine displacement. Routine ASA monitors were applied, and rapid sequence induction was commenced following adequate preoxygenation. Uterine relaxation was initiated and maintained with 2.5 - 3 MAC isoflurane prior to hysterotomy. Maternal hemodynamics was controlled with intravenous ephedrine and phenylephrine boluses as required. Following delivery of baby's head and right arm, a pulse oximeter was applied to obtain preductal oxygen saturations. The anesthesiologist dedicated to baby's care ensured fetal anesthesia and analgesia by administering IM fentanyl and pancuronium. The baby was intubated with a #3.0 endotracheal tube. Attempts to pass a 10 Fr Repogle into the stomach by the surgeons proved unsuccessful. Over the 20- minute trial of gentle ventilation, the baby's oxygen saturations barely rose to the 80s while still on placental bypass and the decision was made to place the baby on ECMO. A peripheral intravenous catheter was placed in baby's left hand through which 50U/kg Heparin was administered. Venous-arterial ECMO initiated and the baby was delivered and umbilical cord clamped. The baby was subsequently transported to the Neonatal intensive care unit. Maternal uterine contraction was initiated with 10U of oxytocin in 500cc LR administered as an IV infusion and supplemented with IM methergine and 10U oxytocin administered directly into the myometrium. Mom was extubated at the end of procedure and was transferred to the Labor & Delivery recovery suite. The baby underwent surgical repair of the diaphragmatic hernia 24 hours later while still on ECMO.

DISCUSSION: Uterine relaxation is the single most important factor that affects the overall success of the EXIT procedure. Consistent uterine relaxation during uteroplacental support to the fetus is necessary to preserve maternal-fetal gas exchange at the placental interface, ensure fetal oxygenation and avoid potentially life-threatening hypoxemia¹. Coordination of care between the anesthesiologists and the surgeon is important to ensure adequate uterine relaxation during uteroplacental support, and prompt contraction prior to delivery of the placenta to prevent exsanguination from uterine sinusoids. The LHR and liver position are important prognostic indicators in fetuses with prenatally diagnosed CDH². A LHR <1 is associated with very high mortality, and liver herniation decreases survival rate by 56% while the survival rate for CDH patients with intraabdominal liver position approaches 100%. The presence of the liver herniation, gastric volvulus and stagnant LHR in this fetus increased the morbidity and potential mortality of this infant. This infant with severe CDH had a high likelihood of requiring ECMO support. Immediate placement on ECMO allows for bypass of the tenuous post delivery course of barotrauma, hypoxia, acidosis and hemodynamic instability. The EXIT to ECMO procedure is associated with favorable survival rates and acceptable pulmonary morbidity in fetuses that would otherwise have an unfavorable outcome with conventional management³. The importance of a multidisciplinary, dedicated team of physicians caring for both fetus and the

pregnant mother as well as a coordinated pre-event planning can not be overemphasized in the successful execution of this treatment strategy.

References:

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