Seminars in Pediatric Surgery 22 (2013) 44–49

Contents lists available at SciVerse ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

# Ex Utero Intrapartum Therapy

Julie S. Moldenhauer, MD*

*The Center for Fetal Diagnosis and Treatment, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania*

ARTICLE INFO

*Keywords:*
Congenital cystic adenomatoid malformation
Congenital diaphragmatic hernia
Congenital High Airway Obstruction Syndrome
Ex Utero Intrapartum Therapy (EXIT)
Fetal neck mass
Placental bypass

ABSTRACT

The Ex Utero Intrapartum Therapy (EXIT) procedure was initially developed to secure the airway in fetuses at delivery after they had undergone *in utero* tracheal occlusion for congenital diaphragmatic hernia. Indications for the EXIT procedure have been expanded to include any delivery in which prenatal diagnosis is concerned for neonatal airway compromise, such as large neck masses and Congenital High Airway Obstruction Syndrome, or when a difficult resuscitation is anticipated such as with large lung lesions. Uteroplacental blood flow and gas exchange are maintained through the use of inhalational anesthetics to allow optimal uterine relaxation with partial delivery of the fetus and amnioinfusion to sustain uterine distension. Using the EXIT procedure, sufficient time is provided on placental bypass to perform life-saving procedures such as bronchoscopy, laryngoscopy, endotracheal intubation, tracheostomy, cannulation for extracorporeal membrane oxygenation, and resection of lung masses or resection of neck masses in a controlled setting, thus avoiding a potential catastrophe.



## Introduction

Prenatal imaging has allowed accurate diagnosis of fetal anomalies that can impact the neonatal airway at delivery. The early recognition of potential airway compromise permits alterations in delivery management, such as ensuring the presence of neonatal resuscitation teams that can be life-saving. Cesarean delivery to facilitate fetal/neonatal resuscitation on placental support was described in early reports to manage airway obstruction.[1,2] The utility of halothane to promote uterine relaxation for fetal tracheal intubation on uteroplacental circulation has also been described and highlighted the need for prolonged uterine relaxation.[3] The EXIT procedure was initially designed to reverse tracheal occlusion performed on fetuses with severe congenital diaphragmatic hernia with additional indications described shortly thereafter.[4–6] With the EXIT procedure, uteroplacental blood flow and gas exchange are maintained through the use of inhalational anesthetics to allow optimal uterine relaxation with partial delivery of the fetus and amnioinfusion to sustain uterine distension. This additional time on placental bypass is used to perform life-saving procedures such as bronchoscopy, laryngoscopy, tracheostomy, cannulation for extracorporeal membrane oxygenation, resection of lung masses, or resection of neck masses in a controlled setting, thus converting a potential catastrophic emergency into a planned procedure. This review is based on our clinical experience with more than 90 EXIT procedures at the Center for Fetal Diagnosis and Treatment of the Children's Hospital of Philadelphia.

## Technical aspects of the EXIT procedure

### *Prenatal evaluation*

Accurate prenatal diagnosis is essential to identify any maternal/fetal pair that are potential candidates for the EXIT procedure. Making the diagnosis early in gestation is also important so that the mother can be referred to a tertiary care center that is not only capable of performing an EXIT, but also has in place the detailed imaging studies that are crucial for perioperative planning. Optimal imaging includes high resolution fetal sonography, three dimensional fetal sonography, ultrafast fetal magnetic resonance imaging, and fetal echocardiography. Amniocentesis for fetal karyotype is recommended. Additional genetic studies may be warranted depending on the presence of associated findings. Care must be taken to assess the placenta, as abnormal placentation such as placenta previa or evidence of subchorionic hemorrhage might increase the risk of intraoperative complications. Proper planning and execution of an EXIT procedure involves a very large multidisciplinary team as outlined in Table 1. The team is also responsible for providing patient education and ensuring parental understanding of the risks, benefits, and potential outcomes associated with the EXIT procedure.

## Procedure

The EXIT procedure is not simply a "fancy" cesarean delivery. It involves the use of general anesthesia, prolonged partial delivery of the fetus, and a large multidisciplinary skilled team. In contrast, cesarean delivery is most commonly performed using regional anesthetic with complete and rapid delivery of the

* Corresponding author at: Julie S. Moldenhauer, MD, The Center for Fetal Diagnosis and Treatment, Children's Hospital of Philadelphia, 34th St & Civic Center Boulevard, Philadelphia, PA 19104-4399.
*E-mail address:* moldenhauerj@email.chop.edu

1055-8586/$ - see front matter © 2013 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1053/j.sempedsurg.2012.10.008

**Table 1**
The EXIT team.

| |
|---|
| Preoperative evaluation: |
| High risk obstetricians |
| Pediatric surgeons |
| Social workers |
| Anesthesiologists |
| Radiologists |
| Fetal cardiologists |
| Neonatologists |
| Intraoperative team: |
| Maternal anesthesiologist |
| Fetal anesthesiologist |
| Scrub nurses[2] |
| Circulating nurses[2] |
| Airway cart nurse |
| Maternal-fetal medicine specialists |
| Fetal cardiologist for intraoperative echocardiography |
| Pediatric surgeons |
| Obstetrician |
| ECMO team |
| Neonatal team: |
| Neonatologist |
| Surgical advanced practice nurses |
| Neonatal nurses |
| Second operating room team on standby in adjacent OR |

neonate, and a much smaller surgical team. The goal of the EXIT procedure is to preserve uteroplacental blood flow for a prolonged period of time so that a surgical procedure can be safely performed on the fetus. This is achieved through uterine relaxation and maintenance of uterine volume. High concentrations of inhalational anesthetics in conjunction with additional tocolytics are used to provide uterine relaxation. In order to maintain uterine volume, only the head and upper torso are delivered onto the maternal abdomen (Figure 1). The lost amniotic fluid is replaced with a continuous warmed amnioinfusion of physiological solution to maintain uterine volume and prevent placental separation (Table 2).

Details of the EXIT procedure have been described elsewhere.[5–8] In brief, the patient is positioned on the operating table in the supine position with a leftward tilt to displace her uterus off of her underlying vena cava, improving preload and avoiding hypotension. Pneumatic compression boots are placed.

**Fig. 1.** Overview of the EXIT procedure. The head and upper torso of the fetus are delivered while the abdomen and legs remain within the uterus. Amnioinfusion catheter is in place. Team members are simultaneously positioning the fetus and placing an IV in the right arm. Pulse oximetry covered by aluminum foil has been placed on the left hand. An airway has been established and the endotracheal tube is in place, prior to initiating resection of the fetal lung lesion.

Preoperative ultrasound is performed to confirm fetal position and placental margins. This allows for planning the approach to laparotomy and hysterotomy. Maternal monitoring consists of continuous electrocardiography, pulse oximetry, invasive arterial blood pressure monitoring, and end-tidal $CO_2$ monitoring. Anesthesia is induced through rapid sequence technique using an intravenous combination of thiopental, succinylcholine and fentanyl. This is immediately followed by endotracheal intubation. Anesthesia is maintained using desflurane, halothane, or isoflurane, alone or in combination, titrated to maintain uterine relaxation. The laparotomy is performed via a low transverse abdominal skin incision. Placental location and planned approach to hysterotomy dictate whether the laparotomy will include muscle dividing or midline fascial entry. Once the uterus is exposed, sterile intraoperative ultrasound is performed to confirm fetal position and map the margins of the placenta. Special considerations to investigate prior to the hysterotomy include breech presentation requiring cephalic version, anterior placenta without an adequate placenta-free window necessitating posterior or fundal uterine entry, or amnioreduction in the presence of massive polyhydramnios to prevent rapid decompression of the uterus and more accurately delineate placental margins. Needle aspiration of large cystic lesions or fetal ascites can also be performed to assist in the manipulation of the fetus. The hysterotomy is typically made in the lower uterine segment in a transverse fashion. However, this is commonly extended into the more muscular, active portion of the uterus. If this occurs, the mother should be counseled to avoid labor in any subsequent pregnancy and anticipate scheduled cesarean delivery. Two full thickness stay sutures are placed under continuous ultrasound guidance in a placenta free portion of the uterus devoid of fetal parts or umbilical cord, and the amniotic cavity is entered using the Bovie. A uterine stapling device is used to create a bloodless hysterotomy as illustrated in Figure 2.[9] The hysterotomy is extended to adequately and easily deliver the fetal head, neck and upper torso without tension on the hysterotomy incision that would cause bleeding. In cases of large neck or oral masses, the hysterotomy might be quite large to accommodate delivery without interrupting the hemostatic stapled hysterotomy edges.

The fetus is then partially delivered up to the level just above the umbilicus and appropriately positioned for further procedures. An amnioinfusion catheter is placed in the uterus and warmed physiologic solution is continuously infused to maintain uterine volume. Intramuscular vecuronium, fentanyl and atropine are given to the fetus to ensure adequate anesthesia. Peripheral

**Table 2**
Essential elements of the EXIT procedure.

| |
|---|
| Uterine relaxation |
| Inhalational agents |
| Tocolytic agents |
| Maintenance of uterine volume |
| Amnioinfusion |
| Partial delivery |
| Fetal |
| Anesthetic |
| Blood product availability |
| Echocardiography |
| Intravenous access |
| Pulse oximetry |
| Maternal |
| Arterial line |
| Blood product availability |
| ECG |
| End-tidal $CO_2$ |
| Pulse oximetry |