Title: EXIT Procedure: Management of a Prenatally Diagnosed, Tracheal Obstructing Teratoma From Birth to Discharge

Moderators: Glenn Mann MD, Michael Lamm DO

Institution: The Children’s Hospital and Montefiore Medical Center. University Hospital for Albert Einstein College of Medicine.

Objectives:
- Explain what the EXIT procedure is and identify the indications for the procedure
- Discuss the unique physiologic implications of the EXIT procedure for the mother and fetus
- Define the anesthetic issues surrounding the EXIT procedure
- Appreciate the interdisciplinary approach to an EXIT procedure and the importance of algorithms and troubleshooting

Case History:
A 37 week and 3 day fetus, with a prenatally diagnosed, large, complex neck mass that is high risk for total airway collapse, is scheduled for delivery via ex utero intra-partum treatment or EXIT procedure in a healthy 25y/o G2P0010.

Questions:
What is the time course for diagnosis and management of a fetus with suspected airway compromise? What is the EXIT procedure? What is the history behind the EXIT procedure? How does it differ from a caesarean section?

Case History:
The prenatal course was otherwise uneventful, without other known congenital defects appreciated. One week prior to planned procedure, a maternal abdominal MRI reveals an exophytic, fluid and solid filled mass, from the anterior neck measuring 6.3x8.1x6.7cm, extending just below the jaw to the upper chest of the fetus.

Questions:
What are the indications for the EXIT procedure? Why is a multidisciplinary approach necessary? Who should be part of this team? What issues need to be discussed with the patient and family? What are the crucial steps to providing a safe, balanced, and successful anesthetic for the EXIT procedure? What contingencies and algorithms should be planned for the delivery?
Case History:
An interdisciplinary meeting consisting of the obstetricians, maternal fetal medicine, neonatal intensivists, obstetric and pediatric anesthesia, otolaryngologists and pediatric surgeons took place five days before surgery to formulate a plan and discuss specifics regarding the delivery. The day before the delivery, a simulation of the EXIT procedure was performed and all contingencies were rehearsed.

Questions:
What is your anesthetic plan for the mother? What are the anesthetic and intraoperative concerns for the mother? What monitoring equipment should be used for the mom? Why is a MAC of 2 or greater needed? What are the hemodynamic concerns with a MAC of 2 for the mother? Are there other medications that could be used to provide uterine relaxation?

Intraoperative care:
The mother underwent general endotracheal anesthesia for EXIT procedure. Prior to induction she received fentanyl 25 micrograms and morphine 0.2 milligrams intrathecally and an arterial line was placed. She was pre-oxygenated and received propofol 200 milligrams and succinylcholine 140 milligrams on induction. She was maintained on rocuronium and initially 2% sevoflurane. Sevoflurane 4.5% was given for hysterotomy and maintained while the fetal head, left shoulder and arm were delivered and the airway was secured.

Questions:
Why do we need to have an estimated fetal weight before delivery? What monitoring equipment should be used for the fetus? What are the anesthetic concerns for the fetus? What anesthetics cross the placenta? What resuscitation medication should be available for the fetus?

Intraoperative Care:
A pulse oximeter was placed on the left hand of the fetus. Fentanyl 50 micrograms, vecuronium 0.7 milligrams, and atropine 80 micrograms were given intramuscularly to the fetus (estimated fetal weight 3.8 kilograms). After suctioning the oropharynx, direct laryngoscopy was performed by ENT and the epiglottis and larynx was easily visualized. A 3.5 uncuffed endotracheal tube was placed and a telescope was passed through the endotracheal tube to visualize the trachea. Severe tracheomalacia was noted. Using a combination of a rigid bronchoscope and flexible bronchoscope through the lumen of the ETT, time was taken to identify the carina and secure an optimal position for the ETT approximately 1cm above the carina. Aspiration of 20 mL of serosanguinous fluid from the complex neck mass was done to take pressure off the trachea. The ETT was secured and the fetus was delivered. Uteroplacental circulation was maintained for 45 minutes. Fetal heart rate remained in the 160’s and oxygen saturations between 30-60% by pulse oximetry throughout the EXIT procedure.
Questions:
What is the role of the pediatric anesthesiologist and neonatal intensivist while the airway is being accessed? What steps need to be taken before the umbilical cord is clamped? What immediate concerns are there for the mother after umbilical cord clamping? Why is coordination with the obstetrician important? Should the mother be extubated? Where should the mother recover?

Intraoperative care:
The umbilical cord was clamped and the inhalational anesthetics were discontinued to help restore uterine tone. The mother received oxytocin, carboprost and methergine to achieve adequate uterine tone and hemostasis. Three units of packed red blood cells were also given for a maternal hematocrit of 22%. Post-delivery the neonate was given to the neonatal intensivists, intravenous access was secured and the baby was resuscitated, stabilized and mechanically ventilated. At the end of the case the mother was extubated without incident.

Post-operative care:
On day of life three the patient was brought to the operating room for surgical removal of the complex mass abutting the great vessels and trachea. An interdisciplinary team consisting of cardiothoracic surgery, otolaryngology and pediatric anesthesia were present. IV access consisted of a umbilical vein catheter, two 24G peripheral IV’s and a radial arterial line. Blood products were immediately available. Anesthesia was maintained with intravenous fentanyl, vecuronium and sevoflurane. The baby had an uneventful operative course and was transported back to the NICU intubated.

Questions:
When should resection of the neck mass occur? What are the intraoperative concerns with tumor resection?

Post-operative course progression:
On post-operative day 4 the patient was brought to the operating room for bronchoscopy and extubation. Bronchoscopy revealed mild tracheomalacia. The patient was extubated and returned to NICU for airway monitoring. The baby was discharged home with the family on day of life 8.

Discussion:
The EXIT Procedure, first introduced in 1989, was developed as a means to reverse tracheal clippings performed in utero on a fetus diagnosed with congenital diaphragmatic hernia. It was subsequently implicated in the use of establishing a neonatal airway in fetuses with CCAM, tracheal atresia, congenital high airway obstruction syndrome, large neck masses and as
a temporizing measure for fetuses needing ECMO before placental separation, as in the case of severe pulmonary and cardiac anomalies.

The EXIT procedure is a special technique, wherein the fetus is partially delivered (head, shoulder and single upper extremity) through a classic or modified hysterotomy, leaving the abdomen, lower extremities, and umbilical cord in utero. This technique utilizes the preserved uteroplacental blood flow as a temporizing measure to maintain adequate oxygenation to the fetus, until a secure airway can be established for the fetus in the extra-uterine environment. Treatment strategies include securing the airway via direct laryngoscopy, rigid or flexible bronchoscopy, tracheostomy, or in rare instances, tumor resection when indicated or institution of ECMO prior to complete delivery.

Pre-operative assessment needs to be thorough and a team approach is crucial to a successful outcome. Newer technologies such as high definition fetal ultrasound, low interference MRI techniques, pulmonary doppler ultrasound, amnioreduction, and chromosomal mapping, have all contributed to the earlier detection, diagnoses, treatment, and preparation of these fetuses’ for the EXIT procedure and transition to extra-uterine survival. Algorithms need to be developed between anesthesiologists, obstetricians, pediatric surgeons and otolaryngologists so that a stepwise approach to securing a viable airway may be conducted during the crucial, time sensitive portion of the procedure. Additionally, the mother and family of the unborn child need to be fully aware of the risks and complication as early on as possible, with discussion surrounding the EXIT procedure performed well in advance.

The anesthesiologist’s role in this procedure is two-fold, and employs the resources of both an obstetric and pediatric anesthesiologist. The obstetric anesthesiologist is responsible for providing a balance between adequate surgical conditions for fetal surgery and wellbeing, while maintaining hemodynamic and physiologic stability in the mother. The role of the pediatric anesthesiologist is to aid the otolaryngologist in securing the fetal airway, maintaining hemodynamic stability and oxygenation in the fetus, administering adjuvant anesthetic agents to the fetus, and preparing for emergency resuscitative measures in the event of fetal distress.

Preparation for a successful anesthetic include detailed pre-natal and maternal past medical history, thorough airway exam, pre-medication with acid neutralizers and anti-emetics, rapid sequence induction and smaller endotracheal tubes. Large bore IV access or central line and arterial line for hemodynamic monitoring should be placed. Blood should be in the operating room for both mother and fetus.

Intraoperative considerations include the need for complete uterine relaxation prior to hysterotomy, to prevent placental separation and complete expulsion of the fetus. This is facilitated via high concentrations of volatile anesthetics during GA. This needs to be balanced with maintaining maternal MAP and perfusion pressures, often via boluses of ephedrine or neosynephrine, in order to provide adequate uteroplacental and fetal blood flow. Other important considerations include continuous fetal monitoring, either by continuous ultrasound or pulse oximetry. To guarantee optimal surgical conditions and fetal well-being, IV access may be required for the fetus. Anesthesia for the fetus is maintained via maternal transfer and augmented
by intramuscular dosing of opioid, atropine, and paralytic as needed. At the time of umbilical clamping and full delivery, tocolysis needs to be reversed, by reducing volatile anesthetic concentrations and administration of utero-tonic agents.

In summary, the EXIT procedure is a new and exciting technique used to secure viable airways or guarantee oxygenation in neonates presenting with complex airway problems or congenital anomalies. The anesthesiologist is faced with special challenges in order to provide adequate surgical conditions for the fetus while maintaining maternal hemodynamics. The keys to success for any EXIT procedure, regardless of its indication, include a thorough knowledge of the unique physiologic challenges, good preoperative assessments for both the mother and fetus, maintaining uteroplacental perfusion, and a step by step team approach to the procedure with open communication.

References:


