## <u>Problem-Based Learning Discussion</u> Planning a Safe EXIT: A Multidisciplinary Approach Stem Case and Model Discussion

**Goals**:

After the completion of the PBLD participants will be able to:

- 1. Examine safety risks and concerns unique to fetal surgical interventions.
- 2. Explore challenges of administering anesthesia to a fetus during an EXIT procedure.
- 3. Discuss the significance and implications of a multidisciplinary approach for an EXIT procedure.

Case:

A 35-year old  $G_2P_1$  woman at 36 weeks gestation is scheduled for an elective cesarean section for an ex utero intrapartum treatment (EXIT) procedure. Fetal magnetic resonance imagining (MRI) revealed a large cervical neck mass with significant tracheal obstruction.

What is an EXIT procedure? What fetal diseases are eligible for the EXIT procedure? What are the main anesthetic goals of an EXIT procedure?

What are the medical specialties that are involved in planning and execution of an EXIT procedure? When is the appropriate time to begin planning for these types of cases? How often should this multidisciplinary team meet? What are the possible scenarios that should be planned for? What role does simulation play in the planning of an EXIT procedure?

Should the EXIT procedure be done in one or two operating rooms? What are the advantages and disadvantages to each option? How would you prepare the operating room(s) for an EXIT procedure?

Where should each team member physically be situated in the operating room during this case? Should you allow trainees to participate in these types of cases?

The mother is induced by rapid-sequence followed by endotracheal intubation. The obstetrician immediately makes incision after the ETT is secured, with successful delivery of the fetus' head and both arms.

What monitoring equipment would you want for the fetus at this point? What are the challenges to monitoring the fetus in the surgical field? What drugs, if any, would you want to have prepared for fetal intubation? How do you plan on administering these drugs?

Based on fetal MRI, the cervical neck mass is located lateral to the trachea on the right side with moderate compression of the airway and subclavian vessels. Initial planning to secure the airway involved direct visualization by laryngoscopy.

# Who should be responsible for intubating the fetus (anesthesiologist or ENT surgeon) in the surgical field?

After intramuscular injection of ketamine 3 mg/kg and succinylcholine 2 mg/kg, the attending ENT surgeon performs direct visual laryngoscopy of the fetus and places a 3.0 cuffed endotracheal tube. He verbalizes that he saw the ETT pass through the cords and that the obstetrician may proceed with full delivery of the baby and placenta.

How would you confirm correct placement of the ETT? Would you confirm ETT placement before or after placental separation? What are the disadvantages of EtCO<sub>2</sub> monitoring when the fetus is still receiving placental blood flow?

After delivery, the baby is carried to the back table for assessment by the neonatology team. Intravenous access is obtained and Apgar scores are 7 and 4. The baby appears to be dusky and you are unable to hear bilateral breath sounds.

#### Who should manage the airway at this point (anesthesiologist or neonatologist)?

You decide to pull out the ETT and manually ventilate the baby by mask. The baby's heart rate is 87, and you are unable to get EtCO<sub>2</sub> or visible chest rise. You urgently place an LMA in the baby and are able to see chest rise and EtCO<sub>2</sub>. After about a minute of ventilation via LMA, heart rate is now 162.

What is the next step in order to secure the airway? Would you attempt fiberoptic intubation? Is it appropriate to perform a tracheostomy on the baby? When should ECMO be considered as an option?

#### **Discussion**:

With advances in prenatal imaging, fetal surgery has now become an option for treatment of several neonatal disorders that are detected while the fetus is in utero. Fetal surgery is generally divided into three categories: minimally invasive surgery (without a large hysterotomy), open mid-gestation procedures, and ex utero intrapartum treatment (EXIT).<sup>1</sup> While these categories differ in technique, all of these options have successfully allowed for palliation or correction of a fetal disorder while fetal circulation and uteroplacental support is still intact. With this circulation providing oxygenation to the fetus, interventions can take place before these disease states can lead to further irreversible damage, such as severe hypoxia or mortality.

### **Indications for EXIT**

The EXIT procedure has been successfully used to intervene in several fetal disease states. These include but are not limited to the following: congenital airway obstructions such as tracheal stenosis/atresia, neck masses, pulmonary and mediastinal masses, congenital diaphragmatic hernia, and congenital heart disease.<sup>2</sup> In some instances, the EXIT procedure can be performed in order to transition the infant to extracorporeal oxygenation (ECMO) until further surgical interventions can be done, such as in hypoplastic left heart syndrome with intact atrial septum.<sup>2</sup> Importantly, the EXIT procedure has allowed for longer interventions (up to 2.5 hours) to be performed in a more controlled and optimized environment than was previously possible.

#### **Preparation for an EXIT Procedure**

The planning and preparation for an EXIT procedure involves a multidisciplinary team of practitioners working together over an extended period of time leading up to the procedure itself. Depending on when the fetal disorder is discovered, this can occur over several weeks or months. Furthermore, the lesion being treated will often dictate the team members involved. In its most basic form, however, the team members involved include:

- obstetric anesthesiology team caring for the mother
- obstetric surgery team
- pediatric anesthesiology team caring for the fetus
- pediatric surgery subspecialist
- neonatologist (in the operating room or accepting care of the neonate at the end of the procedure)
- cardiologist to perform echocardiogram for monitoring
- operating room nurses/surgical techs
- radiologist for fetal imaging
- fetal team coordinator

Through regular meetings, this group of practitioners will develop a plan of action for the EXIT procedure. Serial imaging of the fetus will often dictate changes in these plans as the disease process progresses or improves throughout gestation. Importantly, multiple contingency plans need to be discussed as a part of this process, including scenarios for preterm labor or the inability to deliver the baby as expected.

The roles of each individual involved in the case needs to be established as well, since different specialists often feel that they should take responsibility for the care of the neonate. Communication on who will be the primary team for neonatal intubation, obtaining intravenous access, and initiation of resuscitation, if it becomes necessary, must take place early to prevent confusion during the procedure itself. Whether the trainee has a role in these procedures should also be determined. While these can be great learning opportunities for trainees, EXIT procedures are complex and difficult cases that require

strong skill sets. However, trainees can be able assistants with their knowledge of equipment and the environment.

Multiple scenarios can often be solidified through simulation prior to the day of surgery. For a complicated procedure such as EXIT with many different individuals participating in various roles, simulation of the case activity can be used to guide participants about the different roles and how to manage them in the real situation. Even the physical handing off of the baby from the surgical field to another care area is something that can be simulated for better preparation for the real case. This allows for all steps of the procedure and contingency plans to be acted out in order to identify potential problems. Issues such as space, the large number of personnel, their physical location in the room, and equipment needs can all be determined as well.

In many EXIT procedures, the operative procedure takes place in one operating room. Once the baby is fully delivered, then he/she is taken to a second operating room for further management. This allows for more space and less noise for each team to work once the two patients are separated from one another. At times, however, there is no time for transfer to a different location. In anticipation of this, an area in the original operating room needs to be available to meet the needs of the baby after delivery.

Set up of the operating room needs to account for the two patients being cared for. In addition to the normal set up for the obstetrical part of the case, multiple items need to be available on the sterile field for the fetus.<sup>3</sup> These include, at a minimum:

- Pulse oximetry probe (with multiple backup probes available
- Sterile Ambu bag attached to an oxygen source
- End-tidal CO2 indicator
- Equipment for placement of an intravenous catheter, including tourniquet, flush, and tape to secure it
- Fetal medication syringes and resuscitation equipment (see below)
- Predetermined airway equipment, including but not limited to laryngoscope blades, endotracheal tubes, fiberoptic scope, and surgical airway instruments

A bed should be in the operating room to place the baby on for transport to the second room. Appropriate resuscitation and management equipment for the fetus should be in this second room as well, including umbilical catheters, intravenous fluids, and a ventilator.

#### Anesthetic Considerations

In an EXIT procedure, both the mother and the fetus are patients, each of whom requires their own anesthetic and surgical teams.

#### Anesthesia for the Mother

The goal of anesthesia for the mother during an EXIT procedure is to allow for maximal uterine relaxation while maintaining uteroplacental blood flow. This will help to facilitate the delivery of the fetus's head and neck with an intact uteroplacental circulation. In order to achieve this, the mother undergoes a cesarean section. Most commonly, this is done under general anesthesia. Through the use of inhalational agents (at approximately 2-3 MAC), uterine relaxation is achieved in order to decrease the likelihood of contractions and placental disruption.<sup>4-6</sup> Desflurane, which is a more insoluble agent, has a more rapid tissue response as well as allows for faster onset and recovery from anesthesia.<sup>1</sup> Of concern, there is the potential that prolonged exposure to inhalational agents results in fetal hypoxia and acidosis.<sup>7</sup> To decrease these affects, other options for maternal anesthesia have been used, including a total intravenous anesthetic with propofol and remifentanil or epidural anesthesia.<sup>1,8-10</sup> To achieve optimal uterine relaxation in these cases, intravenous nitroglycerine is also often necessary. Potential side effects include hypotension, reflex tachycardia, tachyphylaxis, and headaches in the awake patient.<sup>2</sup>

No matter which anesthetic technique is used, these procedures are not without significant risk to the mother. Retrospective studies and case reports have shown that EXIT procedures are associated with higher estimated blood loss when compared to cesarean delivery (1104 mL vs 883 mL). Despite this, there does not appear to be a significant difference in postoperative hematocrit.<sup>11-12</sup> Furthermore, the level of maternal anesthesia required to achieve the procedural goals can result in significant maternal hypotension, which must be treated appropriately for both the safety of the mother and in order to maintain uteroplacental perfusion. While ephedrine has traditionally been the vasopressor of choice, phenylephrine has been shown to cross the placenta to a lesser extent and does not appear to affect uteroplacental blood flow as was previously thought.<sup>13</sup> Therefore, either agent is appropriate to use.

#### Fetal Considerations

As described above, the fetus has a separate anesthesia team that is dedicated to his/her care. One of the biggest challenges for this team is fetal monitoring prior to and after partial delivery. Monitoring should begin prior to hysterotomy. This can be accomplished by the pediatric cardiologist who is scrubbed into the case with a sterile echocardiogram probe.<sup>2</sup> Visualization of heart rate, cardiac filling, and contractility are important indicators of fetal well-being during a time when monitoring is otherwise difficult. Once the fetus's head, neck and one arm have been delivered, a pulse oximeter probe can be placed around the exposed hand. Often this is more difficult than expected; wet conditions make contact with the hand tenuous, and exposure to ambient light make readings inaccurate. By drying the hand and covering it with a towel or foil, one is able to get invaluable information on both the fetal heart rate and oxygen saturation.<sup>2</sup> Saturation typically is 60-70% until supplemental oxygen is given to the fetus following intubation.<sup>1,14</sup>

Medications are often needed for the fetus, both to improve chances of successful intubation as well as to anesthetize him/her for optimal pain control and comfort. This can be done via an intravenous (IV) catheter or intramuscular (IM) injection. Intravenous

access can be challenging secondary to the vernix, which makes visualization of potential veins and securing of the line difficult.<sup>2</sup> Even if an IV is planned, multiple IM medication doses should be available in case an IV is not obtained. Medications for anesthesia and potential resuscitaton should include:

- Unit doses of atropine, fentanyl, and a muscle relaxant combined in one syringe
- Atropine 0.02 mg/kg
- Epinephrine 1 mcg/kg

Fluids, including albumen 5% and normal saline, should be drawn up in syringes for volume replacement as needed. Blood should also be in the operating room in case of excessive hemorrhage.<sup>3</sup>

#### Intubation of the Fetus

Extensive preparation for the intubation of the fetus will have taken place in the planning and simulation for the EXIT procedure, with multiple contingency plans discussed and decided upon. First and foremost, which provider will be the first to attempt the intubation needs to be determined. This will often depend on the lesion, the anticipated equipment, and institutional preference. The anesthesiologist, the ENT surgeon, or the general pediatric surgeon are all possible choices. Most importantly, the person chosen should be able to provide a full complement of options to secure the airway as needed.<sup>3</sup> This individual should be in sterile attire at the bedside prior to and during delivery of the fetus. Assistants to that person as well as the second individual to attempt intubation (if the first person is unsuccessful) should also be in sterile gown and gloves, ready to step in immediately if needed.

Steps to follow to secure the airway in a fetus with a large neck mass were nicely described by Olutoye and Olutoye in their review article of 2012.<sup>3</sup> These include the following:

- Direct laryngoscopy
- Rigid bronchoscopy with the endoscope preloaded with an endotracheal tube
- Ultrasound-guided localization of the trachea followed by placement of a guidewire to then use in a retrograde technique
- Perform tracheostomy

Confirmation that the airway is secured can occur in one of several ways. Colorimetric change on an end-tidal carbon dioxide detector as well as lung auscultation (with a sterile stethoscope on the field) can confirm that there is ventilation. If there is any doubt, however, flexible bronchoscopy through the endotracheal tube, and ultrasound assessment of bilateral lung movement can also be used. There should be an increase in the fetal saturation to the 90s from the baseline of 60–70% once ventilation is initiated with oxygen.<sup>3</sup> Confirmation of successful intubation from more than one source as well as agreement amongst team members can help reduce error.

Only after everyone is in agreement that intubation and ventilation are appropriate should the fetus be delivered completely and the umbilical cord clamped.<sup>2</sup> The baby should then be taken to a separate area that is predetermined for further management, including intravenous access, warming and other treatments as needed.

If there is loss of the airway at this point, management will again fall to whoever is most comfortable with securing it at this point. Often, this is the anesthesiologist, the neonatologist, or the surgeon. The difficult airway algorithm should be followed. Airway adjuncts such as laryngeal mask airways, fiberoptic scope, and glidescope should all be available for emergencies. ECMO is often difficult in the baby with a large neck mass due to difficulty with neck cannulation. If this is going to be used, discussion of location to cannulate and the risks and benefits should be planned for prior to the procedure.

The planning and execution of an EXIT procedure is a complex process that requires a large team of practitioners working together to be successful. Otherwise simple care procedures, such as standard monitoring, can be extremely difficult in these cases and require ingenuity and flexibility that may not otherwise be used. However, with good communication and careful planning, EXIT procedures can allow for improved neonatal outcomes.

#### **References:**

- 1. Ngamprasertwong P, Vinks AA, Boat A. Update in fetal anesthesia for the ex utero intrapartum treatment (EXIT) procedure. *Int Anesthesiol Clin* 2012 Fall; 50(4):26-40.
- Garcia PJ, Olutoye OO, Ivey RT, Olutoye OA. Case Scenario: Anesthesia for Maternal-Fetal Surgery. The Ex Utero Intrapartum Therapy (EXIT) Procedure. *Anesthesiology* 2011; 114: 1446-52
- 3. Olutoye OO, Olutoye OA. EXIT procedure for fetal neck mass. *Curr Opin Pediatr* 2012 Jun; 24(3):386-93.
- 4. Marwan A, Crombleholme TM. The EXIT procedure: principles, pitfalls, and progress. *Semin Pediatr Surg* 2006; 15: 107–15.
- 5. De Buck F, Deprest J, Van de Velde M. Anesthesia for fetal surgery. *Curr Opin Anaesthesiol* 2008; 21: 293–7.
- 6. Kuczkowski KM. Advances in obstetric anesthesia: anesthesia for fetal intrapartum operations on placental support. *J Anesth* 2007; 21: 243–51.
- 7. Palahniuk RJ, Shnider SM. Maternal and fetal cardiovascular and acid-base changes during halothane and isoflurane anesthesia in the pregnant ewe. *Anesthesiology*. 1974; 41:462–472.
- 8. Van de Velde M, Teunkens A, Kuypers M, et al. General anaesthesia with target controlled infusion of propofol for planned caesarean section: maternal and neonatal effects of a remifentanil-based technique. *Int J Obstet Anesth.* 2004; 13:153–158.
- 9. Thind AS, Turner RJ. In vitro effects of propofol on gravid human myometrium. *Anaesth Intensive Care*. 2008; 36:802–806.
- 10. Boat A, Mahmoud M, Michelfelder EC, et al. Supplementing desflurane with intravenous anesthesia reduces fetal cardiac dysfunction during open fetal surgery. *Paediatr Anaesth*. 2010; 20:748–756.
- 11. Noah MM, Norton ME, Sandberg P, Esakoff T, Farrell J, Albanese CT. Short-term maternal outcomes that are associated with the EXIT procedure, as compared with Cesarean delivery. *Am J Obstet Gynecol* 2002; 186: 773–7.
- 12. Butwick A, Aleshi P, Yamout I. Obstetric hemorrhage during an exit procedure for severe fetal airway obstruction. *Can J Anesth* 2009. 56:437–442

- Lee A, Ngan Kee WD, Gin T. A quantitative, systematic review of randomized controlled trials of ephedrine versus phenylephrine for the management of hypotension during spinal anesthesia for cesarean delivery. *Anesth Analg* 2002. 94:920 6.
- 14. Schwartz DA, Moriarty KP, Tashjian DB, Wool RS, Parker RK, Markenson GR, Rothstein RW, Shah BL, Connelly NR, Courtney RA. Anesthetic management of the exit (ex utero intrapartum treatment) procedure. *J Clin Anesth* 2001; 13:387–91.
- 15. Lazar DA, Olutoye OO, Moise KJ Jr, Ivey RT, Johnson A, Ayres N, Olutoye OA, Rodriguez MA, Cass DL. Ex utero intrapartum treatment procedure for giant neck masses fetal and maternal outcomes. *J Pediatr Surg* 2011 May; 46(5): 817-22.
- *16.* Myers LB, Cohen D, Galinkin J, et al. Anaesthesia for fetal surgery. *Paediatr Anaest.* 2002; 12: 569-78.
- 17. Tran KM, Maxwell LG, Cohen DE, Adamson PC, Moll V, Kurth CD, Galinkin JL. Quantification of serum fentanyl concentrations from umbilical cord blood during ex utero intrapartum therapy. *Anesth Analg* 2012 Jun; 114(6): 1265-7.