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Delivery of an Infant with Airway Compression Due to Cystic Hygroma at 37 Weeks' Gestation Requiring a Multidisciplinary Decision to Use a Combination of *Ex Utero* Intrapartum Treatment (EXIT) and Airway Palliation at Cesarean Section

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Female, 22-year-old

Final Diagnosis: Central line infection • cystic hygroma • pregnancy

Symptoms: Cystic hygroma • neck mass • pregnancy

Medication: —

Clinical Procedure: Cesarean section • *ex utero* intrapartum treatment • fiberoptic bronchoscopy • general anesthesia • laryngoscopy • rigid bronchoscopy • spinal anesthesia • video laryngoscopy

Specialty: Anesthesiology • Obstetrics and Gynecology • Otolaryngology

Objective: Congenital defects/diseases

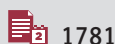
Background: This report describes a case of delivery of an infant with airway compression due to cystic hygroma at 37 weeks' gestation requiring a multidisciplinary decision to use a combination of *ex utero* intrapartum treatment (EXIT) and airway palliation at cesarean section. This infant did not require support with extracorporeal membrane oxygenation (ECMO).

Case Report: A 22-year-old G1P0 woman with past medical history of morbid obesity underwent an EXIT procedure due to a large fetal neck mass. Anesthesia included a narcotic-only single-shot spinal, total intravenous anesthesia (TIVA) was used for maintenance, and high-dose volatile anesthetics and nitroglycerin infusion was used for complete uterine relaxation. The infant's airway was secured by the otolaryngologist, after which delivery was completed. Sevoflurane and nitroglycerin were discontinued and the previous TIVA was restarted. Uterotonics were aggressively administered to prevent uterine atony, and the patient was extubated.

Conclusions: This report shows the importance of a multidisciplinary approach to the management of delivery of infants with airway obstruction. This case demonstrates the approach to the decision for the use of EXIT combined with airway palliation, as ECMO was not combined with EXIT in this case.

MeSH Keywords: Cesarean Section • Extracorporeal Membrane Oxygenation • Intubation, Intratracheal • Lymphangioma, Cystic • Magnetic Resonance Imaging • Obstetric Surgical Procedures

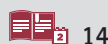
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Background

Morbidity and mortality are significantly elevated in deliveries of neonates with undiagnosed fetal neck masses [1]. Failure to secure the airway in a timely fashion, if at all, in such difficult circumstances leads to extremely poor outcomes for the neonates. The management of delivery of infants with airway obstruction differs in many ways from that of adults and has been detailed by Harless et al. [2]. While ultrasound and magnetic resonance imaging (MRI) allow for superbly detailed examination of these masses, clinically executing the delivery of these neonates remains very challenging and requires extensive collaboration among many teams. The *Ex Utero* Intrapartum Treatment (EXIT) procedure, the successor to the Operation on Placental Support (OOPS) intervention, is a procedure aimed at delivering infants safely that have severely debilitating congenital disorders or other disease processes which otherwise may be fatal. As described in the name of the procedure, the goal is to provide reliable airway treatment externally through the uterus to the fetus prior to delivery [3,4].

The most common indication for the EXIT procedure is congenital airway obstruction, including high airway obstructions and neck masses such as lymphangiomas and teratomas. Other indications include intrathoracic lesions, lung and mediastinal masses, severe bilateral pleural effusions, pulmonary agenesis, diaphragmatic hernias, vascular access in preparation for extracorporeal membrane oxygenation (ECMO), and conjoined twins [1,3,4]. The EXIT procedure was initially designed and performed for the fetus with airway concerns that may prevent survival upon delivery [5]. The fundamental goal is to provide continuous uteroplacental support for the fetus until a definitive airway is secured or a surgical procedure is performed [3,4]. The use of EXIT in a variety of new interventions continues to grow as there are advances in neonatal and obstetrical management. The duration of uteroplacental perfusion continues to be pushed longer and longer, allowing for ever more sophisticated procedures to be performed on the fetus, and thus increasing the chances of survival. According to Rahbar et al., “the fetal gas exchange can be supported by *ex utero* placental circulation for 60 minutes [6].” Also, very importantly, advanced anesthetics are being used in a relatively safe and well-tolerated fashion on the mother of the fetus to avoid increasing the mother’s risk. Maternal anesthetic considerations during the EXIT procedure are numerous, including anxiolysis, pain control, vascular access and advanced hemodynamic monitoring, aspiration and PONV prophylaxis, general versus neuraxial anesthesia, uterine tone, and prevention of post-partum hemorrhage [7]. This report describes a case of delivery of an infant with airway compression due to cystic hygroma at 37 weeks’ gestation requiring a multidisciplinary decision to use a combination of EXIT and airway palliation at cesarean section. This infant did not require support with ECMO.



Figure 1. Delivery of an infant at 37 weeks’ gestation with airway obstruction due to cystic hygroma. The infant is shown at delivery following airway intubation. A large cystic hygroma of the neck is shown, measuring 20.1×9.7×18.2 cm by ultrasound.

Case Report

We present the case of a 22-year-old G1P0 woman with past medical history of morbid obesity (body mass index of 54 kg/m²) who was admitted for an EXIT procedure due to a large fetal neck mass (**Figure 1**) found on 28-week ultrasound. The ultrasound prior to delivery demonstrated a rapidly growing 20.1×9.7×18.2 cm cystic hygroma or lymphangioma, involving the left cheek and extending to the chest and lower abdomen. A fetal magnetic resonance imaging (MRI) study showed a large mass that wrapped around the tracheoesophageal complex, extended into the mediastinum along the left aspect of the trachea, and had a demonstrable mass effect on the mouth, nose, and nasal cavity with rightward displacement (**Figure 2**).

In preparation for the patient’s delivery, a large multidisciplinary team was assembled. The patient had a score of 8 on the Tracheoesophageal Displacement Index, which measures the lateral and ventral displacements of the trachea and esophagus in relation to the cervical spine [8]. The prenatal scans were reviewed at length and there was extensive discussion regarding the most appropriate plan for delivery. Ultimately, it was decided that an EXIT procedure was to be performed due to the potential for severe fetal compromise.

The patient was scheduled for her EXIT procedure at 37 weeks. She received standard obstetrical pre-medications and was taken to the largest operating room in the institution, which in addition to the normal surgical setup, also had a neonatal surgical bed and ECMO circuit immediately available. We administered a narcotic-only spinal block with 100 mcg morphine and 15 mcg fentanyl. The patient was placed in the left uterine displacement of 30 degrees to decrease aortocaval compression and 2 large-bore intravenous lines and an arterial line were placed.

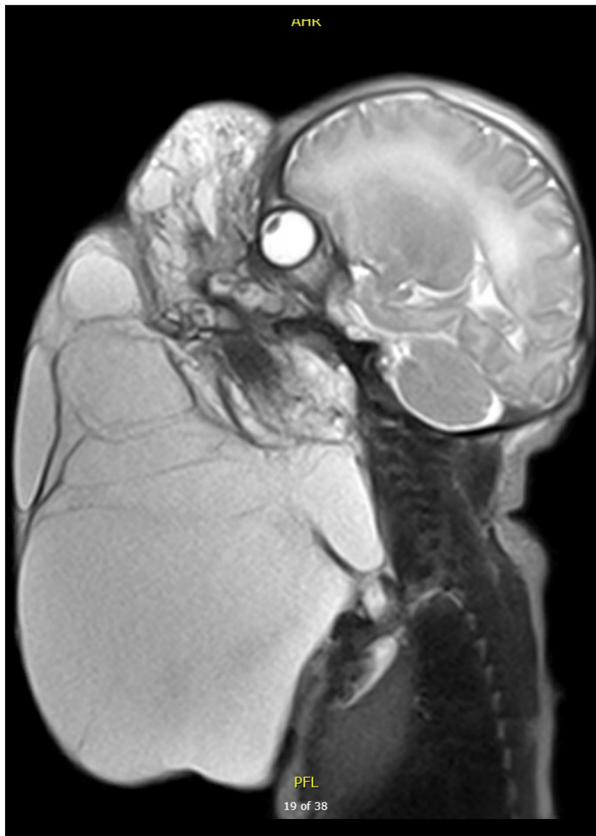


Figure 2. A sagittal magnetic resonance image (MRI) of the infant at 37 weeks' gestation. The MRI shows a large neck mass (left) consistent with a cystic hygroma, measuring 15.8×14×9 cm by MRI.

The patient was induced in rapid-sequence fashion and an endotracheal tube was successfully inserted. Maintenance of anesthesia was via total intravenous anesthesia (TIVA) with 150 mcg/kg/min propofol infusion and 0.1 mcg/kg/min remifentanyl infusion. The patient's mean arterial pressure was kept within 10% of baseline with a phenylephrine titration.

Two minutes prior to hysterotomy, the TIVA was decreased to 25 mcg/kg/min propofol and 2 minimum alveolar concentrations (MAC) of the volatile anesthetic sevoflurane were initiated in addition to a nitroglycerin drip at 100 mcg/min. A small hysterotomy incision was performed and warmed saline was inserted via a Belmont® Rapid Infuser catheter into the uterus. The hysterotomy incision was extended and the infant's head, neck, and right shoulder were delivered, while a sterile warming blanket was placed over the remainder of the uterine incision. The infant received an intramuscular injection of atropine, vecuronium, and fentanyl and the pediatric otolaryngology surgeon began to secure the airway via the oral cavity.

Using many advanced airway tools, the time from initiation of laryngoscopy to securing the airway was 12 min. A combination

of video laryngoscopy, rigid bronchoscopy, and fiberoptic advanced airway equipment were utilized with success, followed by clamping of the umbilical cord and full delivery of a baby boy. Sevoflurane and nitroglycerin were promptly discontinued and full TIVA via propofol and remifentanyl infusions were resumed. The patient received an intravenous bolus of oxytocin, intramuscular carboprost tromethamine, and intramuscular methylergonovine with return of firm uterine tone within 1 min. The remainder of the cesarean section was unremarkable, with 1 L of estimated blood loss and the patient was successfully extubated at the end of the procedure. She was discharged from the hospital on postoperative day 3. The baby underwent debulking and debridement of the cystic neck mass on day 2. Unfortunately, the baby died from fulminant sepsis secondary to femoral central-line infection on day 4.

Discussion

Ex Utero Intrapartum Treatment remains a complex procedure involving significant planning and resources, and it is performed infrequently even at major medical centers across the U.S. EXIT-to-airway, EXIT-to-ECMO, EXIT-to-separation, and EXIT-to-resection are all possibilities depending on the fetal findings on hysterotomy and exposure. Although our case did not require ECMO, a potential failed EXIT-to-airway procedure still necessitated significant planning with pediatric cardiothoracic surgery and perfusionists prior to day of surgery and for these teams to be present with equipment and prepared for emergent EXIT-to-ECMO on the day of surgery. EXIT procedures are of high interest due to their infrequency, high acuity for both mother and infant, and unique anesthetic considerations for uterine tone which changes rapidly during the case.

Anesthesia for an EXIT procedure must focus on both maternal and fetal well-being. Maternal pain control intra- and post-operatively remains a high priority. Options include spinal with intrathecal opioids, CSE with intrathecal or epidural opioids, IV analgesics, and regional anesthetic blocks (e.g., Quadratus Lumborum or Transverse Abdominal Plane blocks). We chose to administer an opioid-only spinal prior to induction as the plan was to induce general anesthesia for patient comfort and anxiety, but also there would be no indwelling catheter in the event of peri-partum hemorrhage and coagulopathy. Institutionally, we routinely remove epidurals prior to leaving the operating room so longer duration analgesia is accomplished with intrathecal morphine. Arterial access remains critical for close maternal blood pressure control to ensure adequate uteroplacental oxygen delivery to the fetus. TIVA using a propofol infusion prior to hysterotomy and after delivery was chosen to help with maternal postoperative nausea and vomiting prevention as the surgery overall was expected to require a long-acting general anesthetic. Remifentanyl, on

the other hand, was used not only for maternal comfort and hemodynamic stability, but has been shown to have transplacental passage, thus helping with fetal analgesia and immobility during surgery or airway manipulation [9].

Uterine tone is an important anesthetic consideration as one must allow maximal uterine relaxation for surgical exposure and fetal airway manipulation with possible ECMO cannulation, but also realizing that strong uterine tone will be critical immediately following delivery to prevent severe post-partum hemorrhage secondary to atony. Therefore, inhaled volatile anesthetics were only used briefly during the surgery for complete relaxation but at a higher MAC than needed for routine surgeries under general anesthesia. Up to 3–4 MAC can accomplish uterine relaxation but with risk of fetal hypoxia and acidosis from decreased uteroplacental blood flow as shown in animal studies if maintained for even 15 min [10]. In addition, nitroglycerin was used for complete uterine relaxation as it has a quick onset and short half-life. Notably, care must be taken with a nitroglycerin infusion as tachyphylaxis can develop, thus requiring higher doses with possible maternal headaches, hypotension, dizziness, pulmonary edema, and methemoglobinemia [11].

Fetal anesthetic considerations include analgesia, fetal monitoring, proper uteroplacental blood flow via tight maternal blood pressure control, maintaining placental attachment, administering intramuscular paralytic and anti-bradycardic medications, and adequate relaxation during airway instrumentation. A Belmont Rapid Infuser® can be used to continuously fill the uterus with crystalloid fluids. This helps with temperature

control inside the uterus, preventing premature placental separation and maintaining the uterine cavity to prevent umbilical cord spasm or compression, which could be detrimental to the fetus [12]. One must be aware that prolonged uterine irrigation with warmed crystalloid can lead to maternal absorption through the venous plexuses and pulmonary edema [13].

Despite careful planning and extensive measures to help fetal survival, newborn deaths following the EXIT procedure are not uncommon. In our case, a central-line infection was the cause of death following a successful EXIT-to-airway procedure. Central-line-associated bloodstream infections account for up to 28 000 deaths per year and considerable costs even with vigilant precautions [14]. More commonly, fetal deaths following EXIT procedure are related to severe pulmonary hypoplasia and immaturity [9]. Maternal outcomes following EXIT appear similar to those of a routine cesarean section but with overall increased blood loss. Ultimately, EXIT is a potentially fetal life-saving procedure that requires a multidisciplinary team, careful planning, and an anesthetic tailored to maternal and fetal comfort and outcomes.

Conclusions

This report has shown the importance of a multidisciplinary approach to the management of delivery of infants with airway obstruction. This case has shown the approach to the decision for the use of EXIT combined with airway palliation, as ECMO was not combined with EXIT in this case.

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