Chapter 11:

ANESTHESIA FOR THE EXIT PROCEDURE

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I. INTRODUCTION

The ex utero intrapartum treatment, or EXIT procedure, was initially described as a method for reversal of tracheal occlusion in fetuses with prenatally diagnosed severe congenital diaphragmatic hernia who had undergone in utero tracheal clip application. Although these infants demonstrated no reduced morbidity compared with conventional treatment options, this novel technique provided a new therapeutic option for fetuses with a variety of potentially fatal diseases. In addition, improvements in prenatal imaging and widespread use of prenatal ultrasound increased the identification of potentially lethal fetal structural malformations, which has had a direct impact on perinatal management and ultimately fetal outcome.

Also referred to as the OOPS procedure (operation on placental support), the EXIT procedure allows controlled delivery and intrapartum assessment strategy to treat fetuses with certain life-threatening diseases. By maintaining uteroplacental circulation with only partial delivery of the infant, crucial time is provided to perform procedures critical to the survival of the infant. These procedures include direct laryngoscopy, bronchoscopy, intubation, tracheostomy, tumor decompression and resection, and extracorporeal membranous oxygenation (ECMO) cannulation prior to clamping the umbilical cord (See Figure 1). In this way, continuous oxygenation is maintained at all times to the threatened infant, thus improving the chances of overall survival. The EXIT procedure is now utilized for infants in which prenatal imaging suggests a very low probability of survival with conventional treatment methods. This group includes fetuses with known tracheal obstruction and other life-threatening airway abnormalities, as well as those who will likely require ECMO support (i.e. congenital cardiac disease and massive congenital diaphragmatic hernia).

Like other types of fetal intervention, the EXIT procedure requires the anesthesiologist to care for two patients at once, often with different and, at times, conflicting requirements. Unlike many other fetal interventions, however, a planned delivery of the infant is the end result of these interventions. This unique difference creates significant increases in maternal morbidity, as these procedures require complete uterine relaxation and serious maternal hemorrhage.
could result. An intimate understanding of the EXIT procedure, the fetal pathophysiology involved, and an understanding of pregnancy-induced alterations directly effecting anesthesia care are required in order to minimize the potentially significant morbidity and mortality of both the mother and her fetus.

This chapter will review the history of the EXIT procedure with particular attention to prior anesthetic management. The fetal diseases most commonly associated with this procedure will also be discussed. Finally, the multiple issues involved in the anesthetic care of both the mother and fetus will be addressed.

II. THE EXIT PROCEDURE: HISTORICAL PERSPECTIVE

The first published report of an EXIT procedure for fetal airway management was in 1989, describing a fetus with complete tracheal obstruction secondary to a prenatally diagnosed cervical teratoma. Intubation was attempted in this premature fetus while maintaining fetoplacental circulation for ten minutes, after which time the fetus’s condition deteriorated and a nonviable infant was delivered. Although this procedure resulted in a poor outcome, the possibility of successful intervention was introduced. Case reports soon followed from various institutions, each describing their experiences with this new technique. Langer stressed the importance of delivering only the upper part of the fetus’s body to better stabilize the fetus through the incision and to avoid manipulation of the umbilical cord. Schwartz described the management of two EXIT procedures: the first performed for a fetus with cervical hemangioma and the second for a fetus with significant epignathus. Tanaka reported a similar experience with a fetus with cystic hygroma, suggesting fetal monitoring with the aid of portable Doppler ultrasound rather than pulse oximetry after encountering difficulties with obtaining continuous fetal heart rate readings during the procedure.

Mychaliska described the first series of eight successfully treated cases using the EXIT procedure and standardized the techniques similar to ones used today. He is also responsible for
introducing the acronym “EXIT” to this intervention. That same year, Liechty and colleagues at the Children’s Hospital of Philadelphia described a systematic approach to the EXIT procedure in a case series of five patients. Of these five patients, two died – one from inability to secure the airway by intubation and no parental permission to perform a tracheostomy, and the second from pulmonary hypoplasia. All cases received preoperative imaging studies, including magnetic resonance imaging with HASTE (half-fourier single-shot turbo spin echo) sequences, which improved the accuracy of diagnoses and provided detail in regards to anatomic relationships. This group also correlated the length of fetoplacental circulation with fetal blood gases, indicating that the fetal oxygenation could be maintained for up to 54 minutes without fetal acidosis. Crucial to the successful outcome of these cases, however, was complete uterine relaxation, obtained primarily with deep inhalational halogenated anesthetic agents. Communication between the surgeon and anesthesiologist, especially during the time of delivery with active management of uterine atony to prevent overwhelming maternal hemorrhage, was emphasized and considered crucial to the overall success of the procedures.

The largest published series to date is by Bouchard and colleagues (2002) who describe their experience with a total of 31 EXIT procedures. In this case series, indications for EXIT procedures were: (1) reversal of tracheal occlusion in patients with a giant fetal neck mass, which accounted for the overwhelming majority of cases, (2) resection of congenital cystic adenomatoid malformation of the lung (CCAM), (3) unilateral pulmonary agenesis, (4) congenital high airway obstruction syndrome (CHAOS), (5) ECMO placement, and (6) aid in separation of conjoined twins. From this series, several guidelines have been suggested. Continuous fetal echocardiography allowed visualization of cardiac hemodynamics, namely heart rate, ventricular volume, and ventricular function, throughout the operative procedures. Although this method required an additional person scrubbed at the operative field, necessitated additional equipment, and demonstrated interference recordings with electrocautery use, it was still considered the most reliable fetal monitor. Pulse oximetry, also used in these cases, was found to be less reliable. Some explanations included an inability to obtain good contact between the probe and the wet fetal extremity, and interference with bright operating room lighting. It should also be remembered that the probes currently used in clinical practice are used to
measure adult hemoglobin, not fetal hemoglobin, making this monitor less reliable. In cases where fetal blood loss was anticipated or in which a prolonged resection was likely, insertion of a fetal intravenous line provided circulatory access for additional analgesia, blood products, and resuscitation medications if needed. As the average maternal blood loss in this series was 848cc, well within the range of that associated with standard cesarean sections, maternal blood administration was rarely needed.

This series was associated with both maternal and fetal complications, however. Maternal complications occurred in two cases. The first resulted from limitations in ultrasonographic identification of the placenta in the face of polyhydramnios. Considerable polyhydramnios may lead to difficulties in identifying the placental edge, which can lead to profound maternal hemorrhage if the placental edge is included in the hysterotomy incision. In one case, the result was a 2.5 liter maternal blood loss. The authors now recommend amnioreduction prior to performing the hysterotomy to map the placental location accurately in cases where significant polyhydramnios exists. The second complication resulted from a prior hysterotomy incision that had dehisced during a subsequent cesarean section delivery with no long-term adverse effects on the mother or baby.

Two fetal complications were also reported in this series. In the first case, inadvertent umbilical cord compression resulted in fetal hemodynamic instability while on uteroplacental gas exchange. The etiology of fetal bradycardia was promptly identified, the umbilical cord position corrected, and one dose on intravenous epinephrine was administered with resolution of the bradycardia. The second fetal complication consisted of a 1mm tear of the trachea during tracheal clip removal. One death occurred in this series, which was a fetus with a large cervical lymphangioma who could not be intubated and whose parents had declined a tracheostomy.

The EXIT procedure has also been performed in twin pregnancies. Leichty described 35 week gestation twins in which one twin had a large neck mass and the other was anatomically normal. The anatomically normal twin was delivered first, and then an EXIT procedure was performed on the twin with the neck mass. The successful outcome of this case was attributed in part to complete uterine relaxation, which not only preserved uteroplacental circulation and fetal gas exchange, but was necessary to perform many of the technical aspects of this case. Midrio reported a case of a twin
pregnancy at 36 weeks gestation in which one twin, located anteriorly, had a large neck mass and the second twin, with normal anatomy, was posterior in location. This group performed an EXIT procedure on the twin with the cervical malformation first, then delivering the normal posterior twin. Both twins had a good outcome. In both cases, the anesthetic management in these cases mirrored the method described by Bouchard et al. No anesthetic complications were reported in either case.

Because of the early experience and outcomes of the EXIT procedure, surgical care has been standardized to some degree, with universal agreement in the need for a team approach, thorough preoperative imaging, careful patient selection criteria, and anesthetic management providing complete uterine relaxation, adequate uteroplacental perfusion, and both maternal and fetal hemodynamic stability. In order to provide superior anesthetic care, however, the intimate details of each fetal disease process must be appreciated, including alterations in fetal physiologic development and potential prenatal treatment options that may very well interfere or alter anesthetic management.

Fetal diseases eligible for the EXIT procedure must demonstrate that without intervention, there would be minimal chance of fetal or newborn survival. Perhaps the best argument for perinatal intervention lies in fetal airway management. Certain fetal airway diseases, when allowed to progress during pregnancy, at best cause significant respiratory distress at birth and, at worst, cause complete airway obstruction incompatible with life. Treatment options prior to the advent of EXIT intervention have resulted in dismal outcomes and high neonatal morbidity and mortality rates. The following section will review the fetal airway diseases most amenable to EXIT intervention.

III. FETAL DISEASES ELIGIBLE FOR THE EXIT PROCEDURE

a. CERVICAL TERATOMA

Cervical teratomas are rare tumors of infancy and represent only 5.5% of all neonatal and childhood teratomas. The incidence is reported to be between 1:20,000 and 1:40,000 live births. Both sexes are affected equally. Congenital cervical teratomas are even more rare, with only 181
cases reported in the literature and 57 cases diagnosed prenatally \(^{11,12}\). Teratomas are composed of tissues from all three germ layers, with neural tissue being the most common component \(^{13}\). These tumors can extend from the mastoid process to the sternal notch inferiorly and to the trapezius muscle posteriorly. They can also invade the oral floor and extend into the anterior mediastinum. Many of the larger teratomas diagnosed prenatally cause maternal polyhydramnios, which is secondary to esophageal compression by the tumor and impaired fetal swallowing \(^{11}\). Most of these tumors are benign at birth but are associated with high mortality rates caused by airway compression and difficulty in establishing an adequate airway after delivery of the infant \(^{14}\). The differential diagnosis includes: congenital goiter and thyroid tumors, cystic hygroma, branchial cleft cyst, neuroblastoma, hemangioma, lipoma, laryngocele, and neural tube defects \(^{11,12}\) (See Table 1).

Because of advances in prenatal magnetic resonance and ultrasound techniques, fetal cervical teratomas are more readily identified, allowing appropriate planning by the medical team prior to delivery of the infant. Fetal MRI is particularly helpful in distinguishing between solid and cystic tissue and tumor components \(^{15}\) (See Figure 2). Even with extensive preoperative preparation, establishment of a surgical airway can still be extremely difficult or even impossible due to anatomical distortion by tumor. Emergent placement onto ECMO may not be possible due to neck and mediastinal tumor involvement. Various attempts to secure the fetal airway have been reported in the literature; some have been successful, whereas others have failed resulting in poor outcomes. Zerella and Finberg reported that 30% of neonates with cervical teratomas and 21% of neonates with cervical teratomas died from airway obstruction shortly after delivery \(^{16}\). Almost all of these reported tumors were benign in nature. For those infants with cervical teratomas that were not diagnosed prenatally, mortality rates are even higher \(^{11,12,17}\). In addition, some larger tumors may interfere with normal delivery methods (i.e. vaginal delivery versus standard cesarean section) and necessitate emergent alterations in maternal care, which places the mother at significantly increased risk \(^{14,18,19}\).

Until recently, treatment options for infants with cervical teratomas who survived the intrauterine period were limited. The standard of care incorporated scheduled cesarean section followed by various airway maneuvers, including establishment of a surgical airway. Even with skilled help immediately available, dismal outcomes were often obtained in those infants with larger teratomas.
due to gross anatomic distortion and difficulty locating the trachea \textsuperscript{11,14,16,17,19} (See Figures 3,4). Even if an airway is established, critical time is needed to perform this task, often at the expense of neonatal oxygenation. With the introduction of the EXIT procedure, precious time is provided to locate the trachea and secure a definitive airway prior to clamping the umbilical cord, thus maintaining continuous fetal oxygenation. These advances have markedly decreased the morbidity and mortality in this subset of patients.

b. Cystic Hygroma

Cystic hygromas are rare causes of prenatally diagnosed neck masses. They arise from the failure of the jugular lymph sacs to join the lymphatic system early in fetal development, resulting in the development of endothelial-lined cystic spaces \textsuperscript{20}. These cystic areas secrete lymph-like fluid, causing enlargement of the cysts and eventual compression of the normal surrounding developing anatomic structures (See Figure 5). This compression may result in fetal hydrops, including fetal skin edema, ascites, and pleural or pericardial effusions \textsuperscript{12,17,20}. The development of hydrops is a poor prognostic indicator \textsuperscript{12,20}. In those patients with isolated cervical cystic hygroma and no evidence of hydrops, airway compromise at birth or shortly thereafter is the main therapeutic concern. It is this subset of patients that are considered candidates for EXIT procedures.

As with cervical teratomas, the true incidence is difficult to calculate, as the numbers of intrauterine deaths, stillborns, and terminations are not included in the literature. The prognosis depends on the fetal age at diagnosis, with earlier gestational age at diagnosis being associated with a worse clinical outcome \textsuperscript{21}. In addition, the presence of prenatal cystic hygroma with other structural anomalies as demonstrated on prenatal sonographic examination is associated with an extremely high mortality rate \textsuperscript{22-24}. These anomalies are not uncommon and include: chromosomal abnormalities, cardiac defects, neural tube defects, hydronephrosis, cleft lip and palate, skeletal abnormalities, imperforate anus, and hydrops fetalis \textsuperscript{20}. Chromosomal abnormalities in particular are found in approximately 60% of fetuses with prenatally diagnosed cystic hygroma \textsuperscript{20,21}. Those fetuses who develop cystic hygromas later in gestation (i.e. third trimester) often present with no other
sonographic indications of anatomic abnormalities nor any evidence of chromosomal defects. As such, these fetuses have a much better chance of postnatal survival provided the fetal airway is not severely compromised at birth.21

Cystic hygromas can also regress spontaneously in utero, presumably due to development of collateral lymphatic and venous systems. Differential diagnosis includes nuchal edema, ecephalocele, cystic cervical teratoma, and twin sac of a blighted ovum.20-24 (See Table 1). The distinction between these diagnoses may be extremely difficult, and every effort should be made to identify any evidence of calcifications, solid components, or bony defects on prenatal sonographic examination that would aid in definitive diagnosis.

c. Congenital High Airway Obstruction Syndrome (CHAOS)

Congenital high airway obstruction syndrome (CHAOS) is a prenatally diagnosed clinical syndrome consisting of extremely large echogenic lungs, flattened or inverted diaphragms, a dilated tracheobronchial tree, ascites, and evidence of nonimmune hydrops.25 The cause of airway obstruction may be due to one of several causes, including laryngeal atresia, laryngeal cyst, or tracheal atresia. Three types of laryngeal atresia are recognized. In Type I, both the supraglottic and infraglottic parts of the larynx are atretic. With Type II, only the infraglottic area is atretic, and with Type III, the affected area involves the glottis.26 Regardless of the etiology of airway obstruction, the clinical presentation is the same. Various anomalies have been associated with CHAOS, although not all patients with CHAOS have associated anomalies.27 These anomalies include: vertebral malformations, esophageal atresia, tracheoesophageal fistula, genitourinary anomalies, imperforate anus, syndactyly, and cardiac malformations. Although the incidence of CHAOS is reported to be very rare, the true incidence may be much more common than originally thought due to intrauterine demise of those affected fetuses. The differential diagnosis for CHAOS is bilateral cystic congenital adenomatoid malformation, which again would be an extremely rare finding.12,25,26,27

Diagnosis of prenatal CHAOS is confirmed by sonographic evidence of complete or near complete upper airway obstruction. Most diagnostic findings result from increased intratracheal pressure and distention of the tracheobronchial tree secondary to the accumulation of fluid in the
lungs. These findings include: uniformly echogenic lungs in the presence of inverted diaphragm, compression of mediastinal structures, and signs of nonimmune hydrops, including fetal ascites, placentomegaly, and pleural or pericardial effusions. Cardiac changes may also been seen, which include the appearance of an elongated heart, septal shift, and small, compressed heart chambers.

Due to the rare occurrence of CHAOS, management guidelines are not definitive. In third trimester fetuses with a diagnosis of CHAOS and no evidence of hydrops, there is most probably incomplete airway obstruction and management is aimed at establishing an airway prior to complete delivery. It is this subset of fetuses who would benefit from an EXIT procedure. Those fetuses with a diagnosis of CHAOS made in the second trimester and those with evidence of complete airway obstruction and/or nonimmune hydrops present a dilemma, as insufficient data exists to determine the best treatment guidelines for this subset of fetuses.

DeCou et al. published the first description of an EXIT procedure performed on a prenatally diagnosed fetus with CHAOS secondary to intrinsic laryngeal obstruction with a favorable outcome. Diagnosis was made at 19 weeks gestation and serial ultrasound exams were performed to monitor fetal well-being. After documenting fetal lung maturity at 35 weeks gestation, an EXIT procedure resulted in successful access to the fetal airway. This infant required respiratory support until diaphragmatic function returned at approximately 8 weeks of life, but subsequently suffered a sudden respiratory arrest and could not be resuscitated. Autopsy revealed a detached fragment of granulation tissue lodged at the carina. Crombleholme and colleagues at The Children’s Hospital of Philadelphia reported the first long-term survivor (age 32 months) of an EXIT procedure for CHAOS diagnosed in the second trimester. This fetus presented at 19 weeks gestation with severe non-immune hydrops, which persisted until an EXIT procedure was performed at 31 weeks gestation. This case suggests that CHAOS diagnosed prior to fetal viability may warrant close observation alone unless there is a pressing need for fetal intervention. Such indications would be rapid progression of hydrops, severe polyhydramnios, or signs of cardiac compromise.
d. **Congenital Goiter**

Fetal goiter refers to a diffuse enlargement of the fetal thyroid gland, which can be associated with fetal hypothyroidism, euthyroidism, or hyperthyroidism. Goiter associated with fetal hypothyroidism is the most common of these entities and is almost always associated with transplacental passage of a thyroid-stimulating IgG antibody from the mother. Such antibodies are present in 90% of women with Grave’s disease. These antibody levels may not reflect maternal thyroid status, making any pregnant woman with Grave’s disease at increased risk for fetal goiter. Other less common etiologies include iodine deficiency, iodine intoxication, congenital metabolic disorders of thyroid hormone synthesis, or hypothalamic-pituitary hypothyroidism. Differential diagnosis depends on whether the fetus is hypothyroid, euthyroid, or hyperthyroid (see Table 1). Sonographic findings of fetal hyperthyroidism include cardiac hypertrophy, tachycardia, or nonimmune hydrops fetalis. Fetal hypothyroidism may be associated with fetal cardiomegaly and heart block. Fetal blood sampling is required to determine the fetal thyroid state.

Regardless of the etiology of fetal goiter, the possibility of significant airway compression immediately after delivery is similar to all fetuses with goiter. In severe cases, even the presence of experienced personnel in the delivery room may not ensure prompt ability to secure an appropriate airway. These patients may benefit from the EXIT procedure, providing the time that may be necessary to identify and secure the compromised fetal airway.

**IV. EXIT to ECMO**

In addition to airway management, the EXIT procedure may be considered for other instances in which separation from uteroplacental support is expected to cause critical cardiac or pulmonary compromise to the fetus. Fetuses with congenital heart disease who are expected to need emergent extracorporeal membrane oxygenation (ECMO) at birth and fetuses with poor prognosis congenital diaphragmatic hernias (liver up, LHR < 1.0) may benefit from the EXIT-ECMO strategy. Patients undergoing this procedure are partially delivered via EXIT, and arterial and venous cannulas are
inserted while maintaining uteropacental perfusion and hence fetal oxygenation. Thus, any period of instability or hypoxia during the neonatal period can be avoided. This is discussed in greater detail in the Chapter 14.

V. **Historical Anesthetic Management of the EXIT Procedure**

While much of the existing literature provides detailed discussions and theories as to the efficacy, surgical technique, and outcomes of the EXIT procedure, very few reports have provided details of the anesthetic management for these interventions. In those few reports that have provided the fine points of anesthetic management, opinions and methods have varied in the recent literature. Most agree that inhaled halogenated anesthetic agents are necessary to obtain complete uterine relaxation. Some have provided additional anesthetic and analgesia medications for the fetus, while some have not. The following section reviews the different techniques reported to date.

Bui *et al* describe an EXIT procedure for congenital high airway obstruction syndrome secondary to laryngeal atresia \(^{40}\). The mother was induced via rapid sequence technique using thiopental, succinylcholine, and fentanyl followed by endotracheal intubation. Isoflurane and nitrous oxide were used, titrating gradually up to an end-expiratory isoflurane concentration of 2.2%. This group used pulse oximetry to monitor fetal heart rate and oxygen saturation. Additional fetal anesthesia was administered intramuscularly to the fetus prior to fetal manipulation with fentanyl 10 ug/kg, vecuronium 0.2 mg/kg, and atropine 5ug/kg. Immediately after delivery, the isoflurane was decreased to aid in return of uterine tone while administering oxytocin as a bolus followed by an infusion. A good outcome was reported with no intraoperative complications. Gaiser and colleagues reported a similar anesthetic technique \(^{41,42}\).

Ward and colleagues report a similar technique, also using isoflurane as the halogenated agent of choice \(^{43}\). This group did report some difficulties, including the lack of specifically designed equipment, sufficient experience, and the inability to stabilize the fetal head due to an "apple bobbing effect". Larsen describe a successful EXIT procedure for a fetus with cervical teratoma with the use of high-dose enflurane, although no details as to concentration used, maternal hemodynamic stability, or fetal perfusion were offered \(^{44}\).
Schwartz et al described the use of sevoflurane to obtain satisfactory uterine relaxation when inspired concentrations from 3% to 6% were used with 50% oxygen and 50% nitrous oxide. The authors concluded that sevoflurane’s lower blood gas partition coefficient, compared with that of isoflurane, allowed faster steady state equilibration. A prior study compared sevoflurane with isoflurane for elective cesarean sections and found no difference in outcomes with low dose 1% sevoflurane versus 0.5% isoflurane for anesthetic maintenance, although the anesthetic goals for an EXIT procedure are quite different than those for a cesarean section. This same group also describes “rapid fetal relaxation” as an advantage of sevoflurane and chose not to give any supplemental fetal drug administration other than intramuscular atropine. The use of a fetal scalp electrode to aid in monitoring fetal heart rate proved to be very helpful in this case report as the intended pulse oximeter did not provide reliable and consistent information.

Not all reported EXIT procedures have used a general anesthetic technique, however. Gagnon et al reports an EXIT procedure for a fetal neck mass performed under an epidural anesthetic technique. This group used magnesium sulfate and nitroglycerin infusions to provide the uterine relaxation necessary to perform the surgery. Although this group recognized the increased risk of prolonged uterine atony and greater maternal circulatory instability with the use of these tocolytic agents, the advantages included an “awake” mother during surgery. This group did not address the amount of vasopressors needed to maintain adequate maternal blood pressure nor did they describe the duration of uteroplacental support. In addition, no fetal anesthesia or analgesia was provided.

The most complete anesthetic management discussion to date was described in the case series previously described by Bouchard and colleagues at the Children’s Hospital of Philadelphia. Anesthesia during the EXIT procedure was maintained with either isoflurane or desflurane. Desflurane was preferred due to its pharmacologic advantages, allowing for rapid decreases in agent concentration in order to regain uterine tone after delivery. Inhalational anesthetic agents provided complete uterine relaxation in all cases and no additional tocolytic agents were necessary. Ephedrine, titrated in 5mg intravenous aliquots, maintained maternal mean arterial pressure at awake values and thus likely assured sufficient uterine blood flow, uteroplacental perfusion, and thus fetal oxygenation. In addition to the placental transfer of inhalation agents to provide fetal anesthesia, fetal analgesia...
and paralysis were further insured with intramuscular fentanyl (10ug/kg) and vecuronium (0.2 mg/kg). Intramuscular atropine (20ug/kg) was also administered to prevent the expected bradycardic response to surgical stimulation often seen in premature infants and neonates. Immediately prior to umbilical cord clamping and delivery of the baby, the inhalational agent was decreased to 0.5 MAC and intravenous oxytocin (20 units in 500 ml) was administered to prevent serious uterine atony and subsequent maternal hemorrhage. Methergine (0.25 mg) and carboprost (250 ug) were available for intramuscular or intrauterine injection if uterine atony persisted. No anesthetic complications were reported.

VI. The EXIT Procedure versus Cesarean Section – The Anesthetic Perspective

The EXIT procedure and cesarean sections are vastly different procedures, each with their advantages and potential risks. Both surgical procedures require specific anesthetic techniques and the ultimate success of both procedures critically depend on a well-planned and executed anesthetic. Although, in both instances, a healthy infant and mother are the ultimate goals, the methods to obtain these end results are vastly different.

Most elective cesarean sections are performed under a regional neuraxial technique (i.e. epidural, spinal, or combined spinal and epidural) with the ultimate goal to deliver a healthy infant as safely and expeditiously as possible, while maintaining maternal safety. As the maternal airway is altered due to physiologic changes during pregnancy, an unrecognized maternal difficult airway should always be considered. Delivery of a vigorous infant involves minimizing medications that, through placental transfer, may have adverse or depressant effects on the newborn. In those cases in which a general anesthetic is employed, concentration of inhalational agents is kept to a minimum in order to avoid respiratory depression in the newborn. A lower MAC will also diminish the risk of uterine atony, thereby minimizing maternal hemorrhage and facilitating placental separation after delivery. However, even in a routine cesarean section, the hysterotomy incision causes alterations in uterine perfusion such that the infant may be at increased risk for hypoxia (See Chapter 3 or 4). In addition, after the uterine incision, continual bleeding occurs from the uterine edges of the incision,
which can lead to significant maternal blood loss and hypotension, which further places the fetus at risk for hypoxia.\textsuperscript{48}

The EXIT procedure, in contrast to a cesarean section, requires complete uterine relaxation prior to hysterotomy. As discussed previously, this is usually accomplished by the use of high dose inhaled halogenated anesthetic agents (2 to 3 MAC isoflurane or desflurane).\textsuperscript{48} Complete relaxation by inhaled anesthetic agents is required for several reasons: (1) surgical manipulation often requires delivering the fetal head, shoulders, and often times an abnormal neck mass through the hysterotomy incision, often not possible with a normal low transverse segment incision used in routine cesarean sections without complete uterine relaxation, (2) the fetus may undergo a surgical procedure and thus will require anesthesia, receives inhalational anesthesia via transplacental transfer, and (3) maintaining fetal oxygenation is dependent on uterine perfusion and minimal uterine vascular resistance. Any alteration in maternal mean arterial pressure or uterine vascular resistance will alter fetal perfusion and ultimately oxygenation.

Complete uterine relaxation places the mother at risk for acute hemorrhage intraoperatively. With the use of a uterine stapling device that seals the membranes and uterine edges during EXIT procedures, the amount of blood loss encountered is markedly reduced. High dose anesthetic requirements also place the mother at risk for hypotension intraoperatively from profound vasodilation, which will in turn cause fetal hypoxia from decreased uterine perfusion if not aggressively and promptly treated. Ephedrine, due to its alpha and beta effects, is the treatment of choice.\textsuperscript{49} Alternative vaspressors include phenylephrine and angiotension II. Angiotension II is thought to cause less vasoconstriction in the uterine vessels and decrease the incidence of fetal academia.\textsuperscript{50,51} In addition, angiotension II does not cross the placenta, placing the fetus at no increased risk from potential side effects.\textsuperscript{52}

Short-term maternal outcomes do not appear to differ between those patients receiving EXIT procedures and those patients undergoing cesarean sections. Scully Noah \textit{et al} compared 38 patients who underwent EXIT procedures to matched cesarean section patients.\textsuperscript{19} Although EXIT procedures took approximately twice as long to perform, there were no statistically significant differences in estimated blood loss, length of hospital stay after delivery, incidence of endometritis,
superficial wound infections or total infections. There were no reported thromboembolic events or maternal deaths in either group in this study.

VII. **Preoperative Considerations**

A. **Imaging Studies**

Preoperative evaluation of the fetal anomaly in question is a crucial step prior to any surgical intervention. Prenatal imaging of all fetal anomalies, including anatomic areas of involvement, relationship to normal structures, and tracheal location are needed to plan the most appropriate surgical intervention. Due to advances in fetal imaging, we are now able to gain a significant amount of useful information. Serial radiologic examinations can also monitor the growth of certain masses, the development of hydrops, and the response to treatment medications (i.e. transplacental digoxin). Hubbard *et al* presented three cases of giant neck masses where MRI aided in prenatal diagnosis and anatomic definition of the mass in relation to the fetal airway. The authors specifically mention the HASTE sequence provided the best definition of a mass because of decreased motion artifacts. Fortunately, these studies can be obtained without any maternal or fetal sedation or fetal muscle paralysis, yet another advantage.

Kalache and colleagues reported the importance of prenatal evaluation of fetal neck masses with the aid of pulmonary Doppler ultrasonography. Fetal breathing movements can be assessed at various levels of the fetal airway such as the trachea and the nasopharynx by combining spectral and color Doppler techniques referred to as pulmonary Doppler ultrasonography (PDU). The authors state that airway compression is a function of the location of the mass, rather than the absolute size of the mass. PDU is useful as this technique examines the fetal airway with dynamic breathing-related information.

Myers *et al* describe the use of combination MRI and Doppler ultrasound to determine potential external anatomic landmarks that may aid in surgical dissection to identify the trachea during EXIT procedures. These studies may be advantageous in certain cases where difficulty in identifying vital structures is anticipated intraoperatively. In this case report, a massive cervical teratoma
completely distorted normal surgical anatomy. With the use of these preoperative studies, combined with intraoperative ultrasound guidance, fetal tracheal rings could be readily identified. The authors suggest this may not have been possible without the use of these combined techniques.

B. Maternal Preoperative Evaluation

Regardless of the proposed surgical intervention, maternal safety is of utmost importance and a thorough medical history and physical examination must be performed in all cases. Included in the physical evaluation is a thorough evaluation of the mother’s airway. Although the physiologic alterations associated with pregnancy are addressed in detail in Chapter 3, any co-existing diseases that could potentially increase maternal morbidity should be addressed. In particular, any cardiac or pulmonary issues that may place the mother at further increased anesthetic risk may render the mother an unsuitable candidate for fetal intervention. Details about the mother’s health, if known, must be obtained. If not available, appropriate studies (electrocardiogram, echocardiogram, pulmonary function tests) should be obtained promptly. In addition, appropriate consultations with specialists will aid in determining the severity of the medical condition(s) and the possibility of stabilizing the condition(s) prior to surgery.

Maternal physiology often reflects the underlying fetal pathophysiology. As most EXIT procedures today are performed for fetal airway obstruction, almost all mothers will have associated polyhydramnios as one of the consequences of fetal airway compromise. A detailed obstetric history including number of amnioreductions, volume withdrawn during amnioreductions, and the presence of uterine contractions associated with these interventions should be obtained. In addition, the presence of baseline uterine contractions is not uncommon with polyhydramnios and this preoperative information is imperative to plan the safest anesthetic technique for both the mother and fetus. A history of preterm labor provides insight into the degree of uterine irritability and usually indicates the need for additional tocolysis during the EXIT procedure. The mother may already be taking oral tocolytic agents or, in some cases, be receiving intravenous tocolytic treatment to control
uterine irritability. Some of these medications (i.e. nifedipine, magnesium sulfate) may alter the anesthetic management significantly (See Chapter 5).

A history of chronic uterine irritability and contractions may indicate a fetus who may be borderline acidotic, thus making this fetus potentially more susceptible to the physiological and pharmacologic alterations associated with the EXIT procedure. The presence of fetal acidemia, regardless of the cause, may necessitate aggressive resuscitation immediately after delivery and may even require the administration of resuscitation medications, including blood products and bicarbonate, to the fetus just prior to commencing with the fetal intervention.

In those cases of fetal goiter necessitating the EXIT procedure, a detailed maternal history and physical examination focusing on exposure to any prior drugs that may affect fetal thyroid function should be noted. These include iodine preparations, propylthiouracil, or amiodarone. The anesthesiologist must also be aware of possible iodine exposure medications such as expectorants or certain radiopaque dyes. A maternal physical examination including clinical signs of hypothyroidism or hyperthyroidism should be performed. Maternal blood analysis, including thyroxine (T4) levels, thyroid-stimulating hormone (TSH) levels, and thyroid-stimulating antibodies should be performed if any concerns arise.

Finally, a complete explanation and description of the surgery itself must be communicated to the mother and her family. Specifically, the patient should have a thorough understanding of the vascular access, necessary monitors, endotracheal and nasogastric tubes, potential side effects of anesthetic medications, and possibility of not seeing her baby immediately after the EXIT procedure. All risks must be completely explained and time must be provided to address all concerns. This interaction should not occur immediately prior to the procedure, but at least 48 hours in advance. In this way, the mother receives the opportunity to privately discuss concerns with her family, which in turn can be addressed well in advance of the procedure.

C. Fetal Preoperative Evaluation

In addition to the preoperative fetal imaging studies previously discussed, an amniocentesis is performed to rule out the possibility of an underlying genetic disorder or other complicating factor that
may not be compatible with extrauterine life. A high resolution fetal echocardiogram should also be performed, paying particular attention to the evidence of fetal nonimmune hydrops, ventricular function, ventricular filling and volume status, and any evidence of fetal arrhythmias. In addition, the ductus arteriosus should be identified and its diameter measured, as the tocolytic agent indomethacin should not be administered if any ductal narrowing is identified. In those fetuses with suspected goiter, evidence of ventricular hypertrophy or bradycardia may indicate the need for pharmacological management with intraamniotic thyroxine prior to the surgical intervention.

Determination of estimated fetal weight is of utmost importance in order to prepare accurate doses for fetal analgesia, muscle relaxation, and resuscitation drugs. Commonly administered drugs include: (1) fentanyl 10ug/kg, given to provide fetal analgesia and to diminish the fetal stress response, (2) atropine 0.1-0.2mg/kg given to prevent the bradycardic response to surgical stimuli, and (3) vecuronium 0.2mg/kg to provide muscle paralysis with a rapid onset of action. Depending on the desired effect, a long-acting muscle relaxant may be given as an alternative (i.e. pancuronium). In those cases where spontaneous fetal respirations are desired soon after the EXIT procedure (i.e. congenital diaphragmatic hernia), a short acting relaxant is desirable to avoid prolonged positive pressure ventilation and possible barotrauma. Cases in which immediate resection of tumor is anticipated would benefit from a longer-acting muscle paralytic agent. Resuscitation drugs (i.e. epinephrine 1-10ug/kg) as well as blood transfusions (irradiated O negative split units) may also be needed in emergency situations.

D. Operating Room Preparation

The hallmark of a good anesthesiologist, regardless of his or her specialty, is thorough preparation for anticipated events before, during, and immediately after a surgical procedure. The ability to predict an event and be prepared for potentially devastating complications associated with a specific surgical procedure can mean the difference between a favorable or devastating outcome. As such, all necessary equipment and drugs should be checked and immediately available if emergently needed. The operating room should be warmed to 85°F in preparation for the partially exposed fetus. Blood products for both mother and fetus should be checked and immediately
available. Blood for potential fetal administration should be in sterile tubing and included in the operative field if needed. The fetal drugs previously mentioned should be in unit doses as calculated by the estimated fetal weight for intramuscular or intravascular administration. These drugs should be prepared in a sterile fashion and ready for immediate administration. Consideration should also be given to the total volume of intramuscular drug administration. We recommend a maximal volume of one ml total, administered in an upper extremity, to achieve results as larger volumes may result in tissue necrosis. Prospective studies are warranted to further delineate the best method for both fetal intravascular and intramuscular drug administration.

VIII. Intraoperative Considerations

A multidisciplinary team consisting of an obstetrician, pediatric surgeon, ultrasonographer, anesthesiologist, neonatologist, and two scrub nurses provide the expertise in each respective field to aid in the overall success of the procedure. In cases where immediate surgical intervention is planned after delivery of the infant (i.e. resection of neck mass), a prepared adjacent operating room with separate personnel should be available. A meeting, usually held prior to the start of the case, identifies individual roles and addresses any concerns or questions the care team may have. This is also a good opportunity to address any clinical changes, either in ultrasonographic findings, change in fetal position, or other factors that may alter the surgical plan.

a. Induction of Anesthesia

Regardless of the anesthetic plan, aspiration precautions should be practiced with administration of 30cc of 0.3 M/L sodium bicitrte to reduce gastric acidity and intravenous metoclopramide to enhance gastric emptying. If a regional or combined regional/general anesthetic technique is to be used, placement of the spinal or epidural should occur after standard ASA monitors are applied to aid in detection of inadvertent intrathecal or intravascular administration of local anesthetic or narcotic. Once the regional anesthetic has been administered, the mother
should be placed in left uterine displacement position. In most cases, general endotracheal
anesthesia is the anesthetic of choice. In this regard, pre-oxygenation is followed by rapid
sequence induction with thiopental (5mg/kg), succinylcholine (1.5-2mg/kg), and fentanyl (1-2ug/kg),
followed by endotracheal intubation. It is important to remember that maternally administered drugs
cross the placenta to reach the fetus immediately. Every maternally administered drug must
be considered in this way prior to delivery as the fetus may exhibit exaggerated responses to these
medications. As discussed in Chapter 3, mucosal edema associated with pregnant maternal
alterations of the maternal airway necessitates the use of a smaller endotracheal tube size (6.0-6.5)
to minimize potential airway complications in the postoperative period.

After the induction of anesthesia, additional intravenous lines, a nasogastric or orogastric tube,
and a temperature probe are usually inserted. Furthermore, a radial arterial catheter is placed in
order to continually monitor of maternal blood pressure as well as provide quick access for lab
determination should serious blood loss occur. As uterine blood flow is directly proportional to
maternal mean arterial pressure, every attempt is made to keep the anesthetized maternal blood
pressure at awake maternal mean values. As discussed in detail in Chapter 4, bolus doses of ephedrine, phenylephrine, or angiotension II, as well as fluid boluses, will maintain
maternal mean arterial pressure with minimal effect on fetal oxygenation.

b. Maintenance of Anesthesia

Prior to maternal skin incision, the inhaled halogenated agent should be increased to 2 MAC in
order to achieve a steady-state equilibrium once intraoperative uterine relaxation is required. It is at
this time that maternal blood pressure should be closely monitored, as decreases in mean arterial
pressure will result in decreased uterine blood flow, decreased umbilical arterial flow, and hence
decreased fetal cardiac output and reduced fetal oxygenation. As discussed in detail in Chapter 4,
Most practitioners recommend the use of 100% oxygen rather than a nitrous oxide or air mixture with an inhaled anesthetic agent of choice. Although this will result in only a modest increase in fetal oxygenation, most agree that the high inhaled anesthetic requirement dictates the use of only agents that will augment uterine relaxation \(^{48}\) (See Chapter 2). Nitrous oxide does not affect the uterine tone to any measurable degree, and thus provides no direct benefit when complete uterine relaxation is desired.

If a general anesthetic is not to be used, other methods must be used to provide adequate uterine relaxation as well as fetal anesthesia and analgesia. Although sparse reports exist in the literature, this can in theory be achieved with intravenous tocolytic agents, such as nitroglycerin boluses or a continuous infusion. Drugs that cross the placental to reach the fetus provide additional analgesia (i.e. fentanyl). This is discussed in greater detail in Chapters 4 and 5.

c. Uterine Relaxation and Perfusion

It is of primary importance to ensure complete uterine relaxation throughout the duration of utero-placental support to the fetus to preserve maternal-fetal gas exchange at the placental interface, ensure fetal oxygenation and avoid potentially life-threatening hypoxemia. Factors affecting uterine blood flow include but are not limited to: anesthetic induction agents, maternal hyperventilation, maternal hypotension, maternal catecholamine release and other etiologies causing increased noradrenergic activity, and increased uterine tone. Uterine blood flow is not autoregulated and is therefore dependent on the maternal mean blood pressure. Any increase in uterine vascular resistance will decrease uterine perfusion, as is seen with uterine contractions (See Chapter 4). Of all factors ensuring the overall success of the EXIT procedure, minimal uterine vascular resistance is the most important as decreases in uterine blood flow will cause fetal hypoxemia, acidosis, and potentially fetal demise.

Although uterine relaxation and perfusion has been extensively discussed in Chapters 4 and 5, it is important to remember that in addition to inhaled anesthetic agents, other tocolytic regimens exist and should be immediately accessible during every procedure.
d. Surgical Procedure

Once the placental location has been confirmed by ultrasound, the maternal abdomen is surgically prepped and draped. An abdominal incision is made and surgical dissection allows for complete exposure of the uterus. Surgical palpation is then performed to assess the degree of uterine relaxation. Although many attempts have been made to develop more subjective means of determining the degree of uterine relaxation, the gold standard to date remains the surgeon's manual assessment of uterine relaxation. As such, the hysterotomy is not attempted until the surgeon determines that complete uterine relaxation has been achieved. In those cases where cystic fluid within a neck mass or massive ascites have precluded delivery through a standard hysterotomy incision, several groups recommend aspiration of the cystic components or ascites prior to the hysterotomy incision. In this way, the necessary size of the incision may be diminished significantly. The hysterotomy location is determined by confirming the borders of the placenta, determining the position of the fetal head and neck, and the umbilical cord with sterile intra-operative ultrasonography. Once the site is confirmed, a stapled hysterotomy is then performed using a hemostatic uterine stapling device (US Surgical Corporation, Norwalk, CT) to minimize maternal hemorrhage from the completely relaxed uterus.

After the hysterotomy site has been created and hemostasis has been achieved, the fetal head, neck and shoulders are delivered into the operative field. As many of these procedures involve large neck masses, a generous hysterotomy is need as well as minimal uterine tone to partially deliver the fetus without injury to the mass or fetus. Furthermore, if a uterine contraction occurs at this time, inadvertent expulsion of the fetus could occur, interrupting the fetal-placental unit and all uteroplacental oxygenation would immediately cease, critically jeopardizing the viability of the fetus. In some cases, a fetal extremity may be delivered in order to apply a pulse oximetry probe and to obtain intravenous access. Although the fetus is anesthetized via placental transfer of maternally administered inhaled anesthesia in most cases, additional analgesia and paralysis are administered (i.e. fentanyl, atropine, muscle relaxant). The additional medications are usually given
as a single dose in an upper extremity or can alternatively be delivered under ultrasound guidance prior to hysterotomy. Advantages to an earlier administration include an increased time for fetal absorption via the intramuscular route. If peripheral intravenous access is obtained, the additional medications can also be given through this route.

e. Access to the fetal airway

Most EXIT procedures are currently performed to access a tentative fetal airway prior to delivery. As such, successful access to the fetal airway is a result of meticulous preoperative evaluation and careful preparation for a variety of intraoperative possibilities. Airway access by means of direct visualization of vocal cords and subsequent endotracheal intubation with conventional laryngoscopes are not usually useful in these cases. Furthermore, portions of the trachea can be completely compressed and distorted such that even successful intubation may result in inability to achieve adequate ventilation. For these reasons, most surgeons perform a direct laryngoscopy and/or rigid bronchoscopy to first examine the status of the fetal airway. In the series from CHOP reporting 77% successful endotracheal intubation, fetal anatomy allowed for endotracheal tube insertion under conventional means (i.e. direct laryngoscopy) \(^3\).

In those cases in which tracheal intubation is impossible, several other options exist. Formal surgical tracheostomy can be performed once the trachea is identified within the surgical incision. The trachea can be located with the aid of preoperative radiographic studies, often identifying the tracheal location relative to fixed external anatomic landmarks. Gentle surgical palpation may also aid in the identification of cartilaginous tracheal rings. In cases in which the former options have failed, the use of ultrasonography with the sterile probe inserted directly into the surgical incision can guide the surgeons to the areas of tracheal rings has also been utilized successfully \(^14\). One must remember that, as in the cases of cervical teratoma, the tumor itself may contain cartilage and bony material, further hampering the success of gaining access to the trachea.

Once tracheal rings are identified, the trachea may be accessed directly with an endotracheal tube, tunneled through the fetal soft tissue, or with the aid of a retrograde wire inserted by the
Seldinger technique. The trachea, exposed through a neck incision, may be incised via a temporary tracheotomy to allow passage of a feeding tube or wire from the trachea to the mouth or nose. The guide-wire is then attached to the endotracheal tube, which is then pulled down into proper position. After suturing the ETT securely to the mouth, the tracheostomy can then be closed primarily with fine absorbable sutures \(^3,^{17},^{32}\)..

Regardless of the method used to secure the trachea, the anesthesiologist must be prepared to control ventilation in the fetus. In some institutions, an anesthesiologist may be scrubbed at the operative field to assume this responsibility. In other institutions, one of the surgeons or neonatologists assumes this role. Adequate ventilation may be difficult to achieve for several reasons. Certain types of tumors, specifically cervical teratomas, may secrete thick mucous into the trachea and this must be aggressively removed with suction prior to ventilation. Once the airway is suctioned with minimal amounts obtained, surfactant should be administered (Infasurf 3ml/kg) into the trachea to diminish the expected airway resistance encountered. These steps should result in increases in fetal oxygen saturation to greater than 90%. If this does not occur, position of the endotracheal tube should be rechecked and the lungs should be auscultated with the aid of a sterile stethoscope. Ventilation occurs most commonly with the aid of a sterile Jackson-Rees circuit with sterile oxygen tubing connected to a reliable oxygen source. Once adequate ventilation has been established, the fetus can be delivered with clamping of the umbilical cord.

**f. Delivery of the Infant and Maternal Management**

Prior to umbilical cord clamping and delivery of the baby, coordination between the surgery and anesthesiology teams is crucial to prevent uterine atony and excessive maternal hemorrhage. As a decrease in the tocolytic agent, whether an inhaled agent or an intravenous agent, would result in increased uterine vascular resistance and decreased fetal oxygenation, reversal of the tocolysis must occur immediately after the umbilical cord is clamped. This is best achieved with low solubility halogenated inhaled anesthetics (i.e.desflurane). As the cord is clamped, the anesthetic agent is immediately discontinued, oxytocin is administered as a bolus followed by a continuous infusion and
titrated to uterine response. Additional uterotonic medications may be necessary and should be immediately available if uncontrolled maternal hemorrhage occurs. These medications include methergine (0.25 mg), carboprost (250 µg), and calcium carbonate (100-200 mg boluses). Blood products should also be immediately available and administered if uncontrolled and persistent bleeding occurs. In cases of uncontrolled hemorrhage despite maximal drug therapy, a hysterectomy may be necessary. It is essential that frequent communication between the surgical and anesthesiology teams continues during this time period, as blood loss can be under-appreciated and the surgeon’s estimate of uterine tone will guide further drug management.

A separate team of neonatologists, anesthesiologists, and nurses should be available for the newborn, as additional medications, blood products, and vascular access may be needed. A brief physical examination, confirmation of bilateral breath sounds, and hemodynamic stability must be insured soon after delivery. In some instances, immediate surgical intervention is planned, necessitating entirely separate anesthetic, surgical and nursing teams in an adjacent operating room as the maternal abdomen is closed.

Once maternal hemostasis has been achieved, uterine tone has been restored, and the placenta delivered, low-dose inhaled anesthetic agents and nitrous oxide can be administered, providing the mother is hemodynamically stable. If an epidural has been inserted for post-operative pain management, it can be bolused with a local anesthetic of choice combined with a narcotic. Many different combinations have been used with success, and it is best to use the combination infusion most commonly used at the home institution. After skin closure, all inhaled agents are discontinued, 100% oxygen is administered, and reversal agents are given. Extubation occurs after the mother is fully awake, follows commands, spontaneously breathing with adequate ventilation, and evidence of protective airway reflexes has returned.

IX. Postoperative Considerations:
Maternal patients recovering from an EXIT procedure differ in some ways from those patients after standard cesarean deliveries. Potential postoperative complications include wound dehiscence, infection, bleeding, and urinary retention, which are similar to those of cesarean sections. Although every attempt is made to place the hysterotomy incision in the lower uterine segment of the uterus during EXIT procedures, those patients with anterior placentas may require incisions in different areas of the uterus. As a result, these patients are at much higher risk of uterine rupture in any subsequent pregnancy. It is therefore recommended that these patients receive cesarean sections in future pregnancies to minimize this risk. In regards to future fertility, a recent study has shown that maternal fertility is not affected by open fetal surgery when the uterine stapling device technique described previously is utilized.

Practitioners should also always consider the fact that, unlike standard cesarean sections, the parents cannot immediately interact or even view their newborn after delivery in many EXIT cases. As many of these newborns undergo immediate surgical intervention, the parents’ first glimpse of their child will be an intubated, sedated child with monitors, invasive catheters, and swollen, distorted facies. Continued emotional support, social services, and education will help ease this transition and provide a positive network for the parents to utilize if needed.

X. Conclusions:

The EXIT procedure has become an important treatment option in the management of certain prenatally diagnosed fetal congenital anomalies. By maintaining the uteroplacental unit, and hence fetal oxygenation, precious time can be provided to perform crucial surgical interventions to ensure oxygenation after clamping of the umbilical cord and delivery. In cases of giant fetal neck masses, these interventions have often proven to be life saving. This procedure can likely be utilized in all fetuses in which serious cardiopulmonary compromise and death are expected after delivery.

The anesthetic management is crucial to the success of the EXIT procedure. Specifically, a thorough understanding of the fetal pathophysiology and these effects on maternal physiology is
paramount to competent anesthetic management. Iatrogenic alterations in uterine perfusion, and hence fetal oxygenation, must be recognized and minimized where possible. Maximizing uterine relaxation is essential to maintain the utero-placental gas exchange. The most important requirement for a successful EXIT procedure, however, is continued communication between all members of the fetal team, including pediatric surgeons, anesthesiologists, obstetricians, neonatologists, and radiologists. It is this well-coordinated team that results in the overall success of these procedures.
REFERENCES:


**Table 1: Differential Diagnosis for the Fetus with Neck Mass**

- Branchial cleft cyst
- Blighted ovum, twin sac
- Cervical meningocele
- Cervical neuroblastoma
- Cervical teratoma
- Congenital goiter
- Cystic hygroma
- Cystic teratoma
- Ectopic thymus
- Hamartoma
- Hemangioma
- Larynocele
- Lipoma
- Lymphangioma
- Neural tube defects
- Neuroblastoma
- Parotid tumor
- Solid thyroid tumors
- Teratoma
- Thyroid cyst
- Thyroglossal duct cyst
Figure 1: Fetal rigid bronchoscopy during the EXIT procedure.
Figure 2:

Fetal MRI, demonstrating massive oropharyngeal teratoma. Note the hyperextension of the fetal neck and upper extremity soft tissue edema, secondary to mass effect from the tumor.
Figure 3:

Fetus with large oropharyngeal teratoma, as viewed immediately after delivery of the head and shoulders during an EXIT procedure.
Figure 4:

Newborn with massive oropharyngeal cervical teratoma, immediately after EXIT procedure was performed to secure the airway. Immediate resection followed in an adjacent operating room.
Figure 5:

Fetus with cystic hygroma who underwent an EXIT procedure in order to establish a surgical airway.