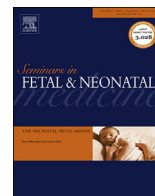




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Review

Immediate operative management of the fetus with airway anomalies resulting from congenital malformations

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Prenatal diagnosis has transformed the outcome of fetuses with airway obstruction. The thorough evaluation of prenatal imaging allows for categorizing fetuses with airway compromise into those who will require a special mode of delivery and those who can be delivered without any special resources. The ex-utero intrapartum treatment (EXIT) approach allows accessing the airway while the fetus is under placental support, converting a potentially catastrophic situation into a controlled one. An expert multidisciplinary team is the key to success.

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1. Introduction

Fetuses with airway obstruction at delivery are at great risk of hypoxic brain injury and mortality. The advent of prenatal diagnostic strategies galvanized the perinatal management of such fetuses. The first example of this was reported by Holinger et al. in 1985. Prenatal diagnosis of a cervical teratoma allowed for special airway equipment to be pre-emptively mobilized to the delivery room prior to cesarean delivery. Direct laryngoscopy and bronchoscopy was used to secure the airway immediately after delivery [1]. The concept of accessing an obstructed airway while keeping the fetus under placental support was first reported by Kelly et al. in 1990, in which direct laryngoscopy was performed to secure the airway [2]. A few months later, Levine et al. reported the sentinel case of a tracheostomy performed on a fetus under placental support in order to secure an airway [3]. No maternal interventions to prevent uterine contractions were performed in these reports, and the fetuses were entirely delivered from the uterus prior to airway manipulation. During the early 1990s this concept evolved until, in 1996, a team at University of California, San Francisco, described a standardized procedure called ex-utero intrapartum treatment (EXIT) [4]. They reported the reversal of a tracheal occlusion in patients with prenatally treated congenital diaphragmatic hernia.

The EXIT procedure is now the gold standard strategy for delivering fetuses with congenital airway obstruction. It permits the fetal airway to be secured in an elective, controlled manner while the fetus remains under placental support. Once adequate ventilation is secured, the fetus can be safely taken off placental support. The indications for an EXIT procedure have grown over time and now include a myriad of conditions, including cervical lymphangiomas, cervical teratomas, large congenital lung malformations, conjoined twins, and congenital high airway obstruction syndrome (CHAOS) [5–7]. Successful performance of an EXIT procedure rests on both thorough preoperative planning and the availability of a highly skilled multidisciplinary team.

2. Prenatal assessment

It is of paramount importance to establish an early diagnosis in order to facilitate a prompt referral of the mother and fetus to a center proficient in performing an EXIT and fetal surgery. The prenatal evaluation of a fetus with airway obstruction includes amniocentesis and other genetic tests, if warranted, but, most importantly, a thorough assessment of the fetal anatomy with fetal three-dimensional ultrasound, fetal echocardiography and ultrafast fetal magnetic resonance imaging (Figs 1 and 2).

3. Team approach and equipment

The EXIT procedure can only be successful if performed by a multidisciplinary team that includes anesthesiologists, pediatric surgeons, neonatologists, maternal–fetal medicine specialists,

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Fig. 1. Cervical teratoma as seen on three-dimensional ultrasound imaging. Reprinted with permission from Lioy J, Sobol S editors. *Disorders of the neonatal airway: fundamentals for practice*, p. 182. © Springer, 2015.

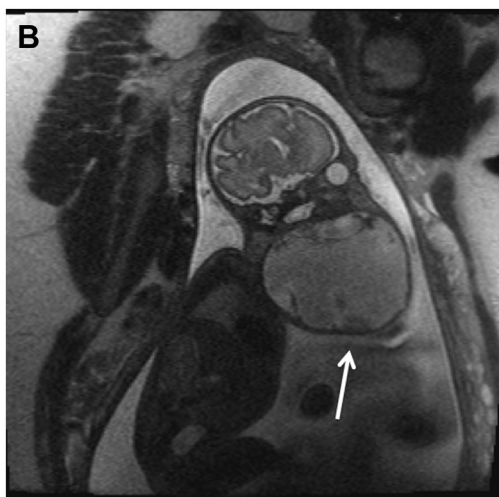


Fig. 2. Ultra-fast prenatal magnetic resonance imaging. (A) Twin pregnancy with one fetus with a cervical teratoma (black arrow). (B) Giant cervical teratoma (white arrow) in a fetus with cystic and solid components. Reprinted with permission from Lioy J, Sobol S editors. *Disorders of the neonatal airway: fundamentals for practice*, p. 182. © Springer, 2015.

fetal cardiologists, and a committed group of operating room nurses and personnel. Extensive preoperative planning, simulations, and ad-hoc team meetings are mandatory. The entire range of airway equipment needs to be sterilely available, including rigid infant-type laryngoscope handles, a set of number 0, 1 and 2 straight Miller blades, rigid ventilating bronchoscopes (sizes 2.5 and 3.0), wire-reinforced cuffless endotracheal tubes (ETTs), tracheostomy tubes, and all surgical instruments required for a neonatal tracheostomy. Once the airway is established, a sterile Mapleson circuit with a manometer and a 1 L bag is used on the sterile surgical field to help ventilate the fetus. Fetuses undergoing an EXIT procedure frequently need immediate post-EXIT surgical interventions for which, ideally, a second operating room must be readily available next to the room where the EXIT takes place.

4. Timing of delivery

Under ideal circumstances, a fetus with airway obstruction should reach term and be electively delivered. However, this may not be possible in some situations as the cause of airway obstruction and its effects on fetal health may dictate otherwise. For example, large cervical tumors may result in compression of the airway and the pharynx/esophagus, resulting in polyhydramnios and medically unresponsive preterm labor. When a mother carrying a fetus with airway obstruction develops signs of impending labor (such as severe recurrent polyhydramnios or shortened uterine cervix) it is crucial to perform an elective preterm EXIT at a time of day when required personnel are available. This is preferable to waiting for the fetus to reach term due to the risk of developing labor at off-hours when the resources necessary for an EXIT may not be easily available. A large published series of fetuses with cervical teratomas having undergone an EXIT reported that only 23% of the fetuses reached term [8].

5. Ex-utero intrapartum treatment

Undoubtedly, deep maternal general anesthesia is essential for a successful EXIT procedure, as general anesthetic drugs are the strongest uterine relaxants currently known and utero-placental gas exchange only happens if the uterus is relaxed. In order to facilitate the management of postoperative maternal pain, an epidural catheter is employed. The mother is meticulously monitored during the EXIT procedure with continuous electrocardiogram, pulse oximetry, invasive monitoring of arterial blood pressure, urinary output with a catheter, core body temperature and peripheral nerve stimulation. The combination of drugs used for induction and maintenance of anesthesia during an EXIT procedure may vary, but the principle of deep inhalational anesthesia must not change. The cornerstone from an anesthesiologist's point of view is to maintain the fetal-placental circulation, which requires adequate uterine relaxation and maintenance of uterine perfusion pressure. Two patients must be anesthetized for the benefit of one, and there is little margin for error [9].

The induction of maternal anesthesia is usually done with intravenous propofol (2 mg/kg) closely followed by a muscle relaxant such as succinylcholine (1 mg/kg) to facilitate a rapid endotracheal intubation. Maintenance of anesthesia is done with desflurane (5–10%) titrated to uterine tone as evaluated by direct palpation during the procedure. Vecuronium (0.1 mg/kg/dose boluses, repeated pro re nata) is used for muscle relaxation. Maternal hypotension may occur as a response to the deep general anesthesia required to facilitate uterine relaxation. This must be

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