

General Anesthesia in an Ex-Utero Intrapartum Treatment (EXIT) Procedure for a Neonate with NAGER Syndrome

Brandon C. Grinage, M.D., Robert O'Bannon, M.D.

Anesthesiology and Perioperative Medicine, Medical College of Georgia, Augusta, Georgia

Case

A 30-year-old G2P1-0-0-1 white female at 38 weeks and 6 days presents for scheduled Ex-Utero Intrapartum Treatment (EXIT) procedure for fetal craniofacial abnormalities secondary to Nager Syndrome. The patient (67 in., 97 kg) has a good airway exam, and a medical history significant only for GERD. Her only current medication is PN vitamins. Labs and vital signs are WNL.

EXIT Procedure

In the EXIT procedure, an incision is made similarly to that for a cesarean section. Following uterine incision, the baby is partially delivered by the obstetrician but remains attached to the maternal-fetal unit, allowing the pediatric surgeon to establish or secure an airway while the baby maintains oxygen saturation on utero-placental support. Once the airway has been secured, the obstetric team resumes control of the procedure, the umbilical cord is cut and clamped, and the delivery of the baby is completed.

The EXIT was initially developed to reverse tracheal occlusion in fetuses who had undergone tracheal clip application in utero for severe diaphragmatic hernia.¹ However, since the first published report in 1989, the role of the EXIT procedure has increased dramatically. Indications now include malformations of the fetal airway, fetal neck and lung masses, congenital goiters, congenital high airway obstruction syndrome (CHAOS), and EXIT to ECMO.

While the EXIT procedure may appear to simply be a cesarean section interrupted by a pediatric surgery, it is a far more complex operation. First and foremost, there must be clear and constant coordination among multiple teams including obstetrics, obstetric anesthesia, pediatric surgery, pediatric anesthesia, and pediatric teams. The anesthetic considerations are far more complicated as well. It is imperative to maintain complete uterine relaxation until the baby is free from utero-placental support to ensure adequate fetal oxygenation while at the same time preventing uterine atony and its resultant maternal hemorrhage.



Our Approach

In the operating room and prior to the case, nurses and physicians discussed logistics and the needs of each team. We went over the position of each team during the case, instrumentation, equipment set-up, placement of fetal monitoring, and individual roles.

Prior to transport to the operating theater, the patient received metoclopramide 10 mg and famotidine 20 mg IV. On arrival to the OR and prior to induction, we placed a right radial artery arterial line. Rapid sequence induction was performed with propofol and succinylcholine. Intubation with a 6.5 mm endotracheal tube was performed under direct laryngoscopy without difficulty.

General inhaled anesthesia was maintained with sevoflurane due to its uterine relaxation effects (vs isoflurane)² as well as its rapid fetal relaxation.³ Shortly after skin incision but before uterine incision, a nitroglycerin infusion (0.5 mcg/kg/min) was begun. The fetus's head and shoulders were delivered and pulse/ox sensor placed on the baby's right hand. ENT then intubated the fetus using rigid bronchoscopy. Time elapsed from partial delivery to intubation was 16 min. After confirming ETT placement, the delivery was completed and the NICU team assumed care of the infant. 5-min Apgar score was 9 (1 point deducted for consciousness). The nitroglycerin infusion was discontinued immediately after the infant was delivered. After the placenta was delivered, pitocin 40 units and methylergonovine were administered to promote uterine contraction and prevent hemorrhage. The mother remained stable, was extubated in the OR, and was discharged home on POD 4 without perioperative complications.

Conclusion

Multiple etiologies in Nager syndrome including craniofacial abnormalities necessitate securement of an airway via the EXIT procedure prior to fetal delivery. Outcomes for both mother and child rely on open communication among all teams and maintenance of the utero-placental unit to provide adequate oxygenation of the fetus prior to delivery.

References

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- ²Schwarz DA, et al. J Clin Anesth 2001;13(5):387-391
- ³Gambling DR, et al. Anesth Analg 1995;81:90-95