An EXIT Procedure in a Patient with Malignant Hyperthermia

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Introduction: The Ex-utero Intrapartum Treatment (EXIT) procedure was initially developed for reversing tracheal occlusion in fetuses with severe congenital diaphragmatic hernia. Since then, the EXIT procedure has expanded to include a bridge for treating everything from airway obstruction to mediastinal tumors to extracorporeal membrane oxygenation (ECMO). We present a case of an EXIT procedure in a fetus with a neck mass, possibly compromising the airway, whose mother has a family history of malignant hyperthermia.

Case Description: The anesthesia machine was prepared for a patient with a history of malignant hyperthermia by removing the inhaled anesthetic vaporizers and running the O2 at 10 liters/min for 20 minutes. A neonatologist and ENT surgeon were present in addition to a separate anesthesia team for the mother and the baby. After placement of standard ASA monitors and an arterial line, a rapid sequence induction was performed with propofol, fentanyl and rocuronium. Following tracheal intubation, a propofol and remifentanil infusion was started at 100 ug/kg/min and 0.1 ug/kg/min, respectively. Approximately 15 minutes after anesthesia induction, skin incision was made and infusions of nitroglycerin at 16 ug/kg/min for uterine tocolysis, and phenylephrine for hemodynamic support were started; the propofol and remifentanil infusions were increased to 175 and 0.25 ug/kg/min, respectively. The fetal head and right shoulder were delivered 13 minutes later. A Miller 1 blade was used for laryngoscopy and the trachea was intubated with a 3.0 uncuffed tracheal tube. A pediatric fiberoptic scope was passed into the tracheal tube to confirm tracheal placement. Six minutes elapsed during these maneuvers during which the fetus appeared anesthetized. APGAR scores were 4 and 9 at one and five minutes, respectively. After delivery of the baby, the nitroglycerin infusion was discontinued and an oxytocin infusion was started. The uterus contracted adequately, and the phenylephrine drip was also discontinued. The mother’s trachea was extubated at the end of the surgery and she recovered without incident. An MRI performed on day 2 of life demonstrated the neck mass was a lymphangioma. The baby was tracheally extubated on day of life 3, and follow-up with ENT is planned.

Discussion: The EXIT procedure has been well described but not in the mother with a family history of MH. Traditionally, a potent inhaled anesthetic agent is administered to anesthetize both the mother and the fetus and to relax the uterus. In this case, due to the MH potential, IV remifentanil and propofol were used as the anesthetic and NTGL for uterine relaxation. These drugs provided excellent anesthesia for the baby and maintained uterine relaxation.