



## Case report

## Ex utero intrapartum treatment procedure for management of congenital high airway obstruction syndrome in a vertex/breech twin gestation

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## ABSTRACT

Congenital high airway obstruction syndrome (CHAOS) is one indication for the ex utero intrapartum treatment (EXIT), which is used to secure the fetal airway, while fetal oxygenation is maintained by uteroplacental circulation. We report a successful EXIT procedure in a twin gestation in which one child had CHAOS while the other was a healthy child without any congenital abnormalities. After version of Twin B to allow for delivery of Twin A, Twin B underwent airway evaluation and tracheostomy for laryngeal atresia prior to delivery.

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### 1. Introduction

Advances in prenatal diagnosis through ultrasound and magnetic resonance imaging (MRI) have led to early detection of fetal airway anomalies, including congenital diaphragmatic hernia, fetal neck mass, and congenital high airway obstruction syndrome (CHAOS). Without intervention, these anomalies can be fatal at birth [1,2]. In the ex utero intrapartum treatment (EXIT) procedure, the fetus is partially delivered, while oxygenation is maintained via the placenta. This technique provides time to secure the neonatal airway utilizing intubation, endoscopy, or surgery, in order to prevent neonatal hypoxemia [3].

Here, we describe a successful EXIT for a twin gestation. Because Twin B was breech, she underwent a successful version before cesarean delivery of Twin A. After Twin A was delivered, the EXIT was carried out for Twin B, who had CHAOS and underwent tracheostomy before completion of delivery.

### 2. Case description

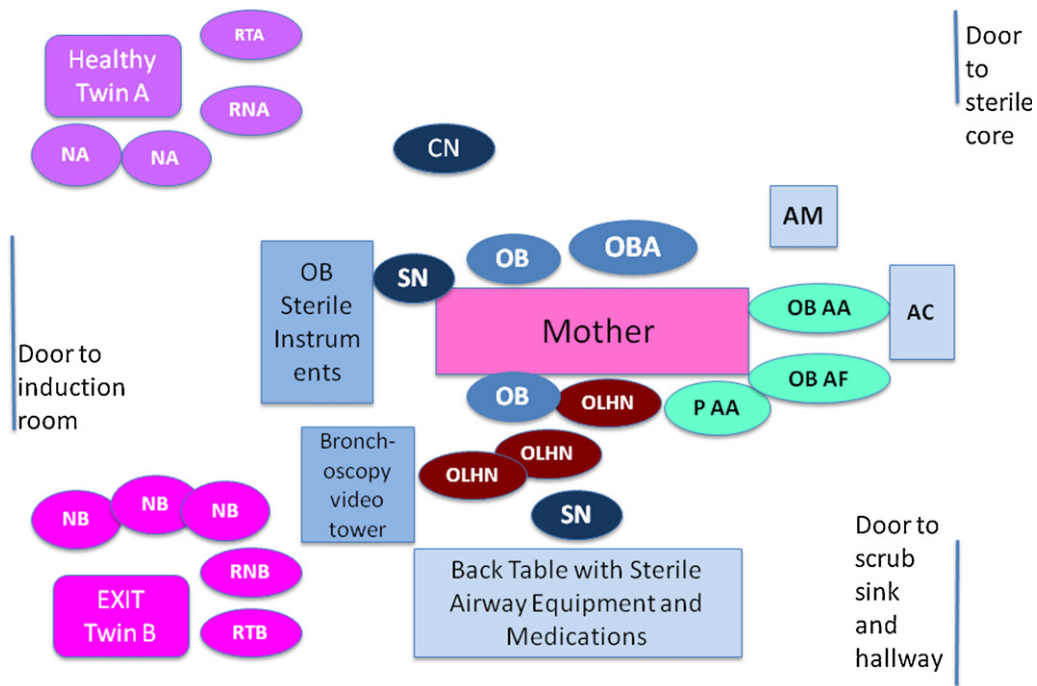
A 44-year-old, gravida 4, para 0 woman with gestational hypertension and a dichorionic, diamniotic twin pregnancy was referred for evaluation by the maternal fetal medicine and otolaryngology groups at 21 weeks 5 days gestation for CHAOS in Twin B; diagnosis was confirmed by ultrasound. After multidisciplinary consultation and discussion with the patient, all parties agreed that performance of the EXIT procedure was the best management option. Weekly sonograms confirmed that fetal hydrops did not develop. Additionally, fetal MRI was performed at 34 weeks and 1 day to better delineate Twin B's fetal anatomy.

Before delivery, a multidisciplinary planning session was held that included teams from Maternal & Fetal Medicine, Pediatric Otolaryngology, Neonatology, and Anesthesiology (Pediatric and Obstetric), as well as operating room (OR) staff. Two days before the EXIT procedure was scheduled, the team conducted a rehearsal with the OR staff (Fig. 1).

The patient was admitted at 36 weeks' gestation. Ultrasound showed a vertex Twin A and a superior, anterior breech Twin B. The surgical plan thus included intraoperative sonogram and intra-abdominal/extruterine version of Twin B to allow for the delivery of Twin A before performance of the EXIT procedure on Twin B. MRI imaging of the congenital high airway obstruction revealed

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**Fig. 1.** Diagram of operating room during EXIT procedure. AC, anesthesia cart; AM, anesthesia machine; NA, neonatal attending for Baby A; NB, neonatal attending for Baby B; OB AA, obstetrical anesthesia attending; OB AF, obstetrical anesthesia fellow; OBA, obstetrician attending; OB, obstetrician resident; OLHN, otolaryngology head and neck resident; OLHNA, otolaryngology head and neck attending; P AA, pediatric anesthesia attending; RNA, nurse for Baby A; RNB, nurse for Baby B; RTA, respiratory therapist for Baby A; RTB, respiratory therapist for Baby B; SN, scrub nurse.

hyperinflation of the lungs with an area of obstruction seen in the upper trachea (Fig. 2).

At 36 1/7 weeks' gestation, the patient was brought to the OR. Before her arrival, a team time-out was called during which all personnel identified themselves and their role. Two anesthesiology teams (one for the mother and one for the baby undergoing airway management), two neonatal intensive care unit (NICU) teams (one for each baby), two pediatric otolaryngologists, and two obstetricians were present (Fig. 1). The case plan was reviewed; equipment, drug supplies, and blood availability were verified. The

patient was brought into the OR and placed in the supine, left uterine displacement position. Standard monitors and external fetal monitors were applied, and the patient was pre-oxygenated. The patient was anesthetized by rapid sequence induction with 100 mg of lidocaine, 200 mg of propofol, and 120 mg of succinylcholine, and then intubated. A second, large-bore peripheral intravenous line and a radial arterial line were subsequently placed. To facilitate uterine relaxation, deep inhalational anesthesia was established with desflurane. Vecuronium was used to maintain muscle relaxation. A nitroglycerin infusion was titrated to achieve additional uterine relaxation, while a phenylephrine infusion was titrated to maintain maternal blood pressure and uteroplacental perfusion. Twenty-six minutes after induction, anesthetic conditions were appropriate to allow the obstetricians to start the surgery.

A vertical skin incision was made and low uterine exposure was achieved. Intra-abdominal/extrauterine version of Twin B was successfully performed. The position of Twin B was maintained by an obstetrician thereafter. Thirty minutes after surgery start time, an incision was made in the uterus while preserving intact membranes. Fetal vertex presentation was reconfirmed, and membranes for Twin A were ruptured. Twin A was delivered without difficulty as a cesarean section through the hysterotomy and was passed to the NICU team A. She was intubated, weighed (2500 g), and transported to the NICU. Apgar scores were 2, 4, and 6 at 1, 5, and 10 minutes, respectively. She was extubated and breathing room air within hours of delivery.

Throughout delivery of Twin A, Twin B was monitored with a sterile ultrasound probe. After delivery of Twin A, Twin B's head was guided to the uterine incision, and the fetus was situated for the EXIT procedure. Once positioned, membranes were ruptured and the head, neck, and right upper extremity were exteriorized. Warmed normal saline was infused into the uterus to maintain uterine volume.

The pediatric anesthesia team administered a single injection of 5 mcg/kg fentanyl, 0.2 mg/kg atropine, and 1.5 mg/kg rocuronium



**Fig. 2.** CHAOS syndrome on prenatal magnetic resonance imaging (MRI). Fetus B with significantly enlarged lungs (A). The most proximal segment of the trachea is not fluid filled (arrow) while the distal 4/5 of the trachea is fluid filled and dilated. There is also a mildly increased amount of amniotic fluid in fetus B with no definite signs of an additional esophageal atresia.

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