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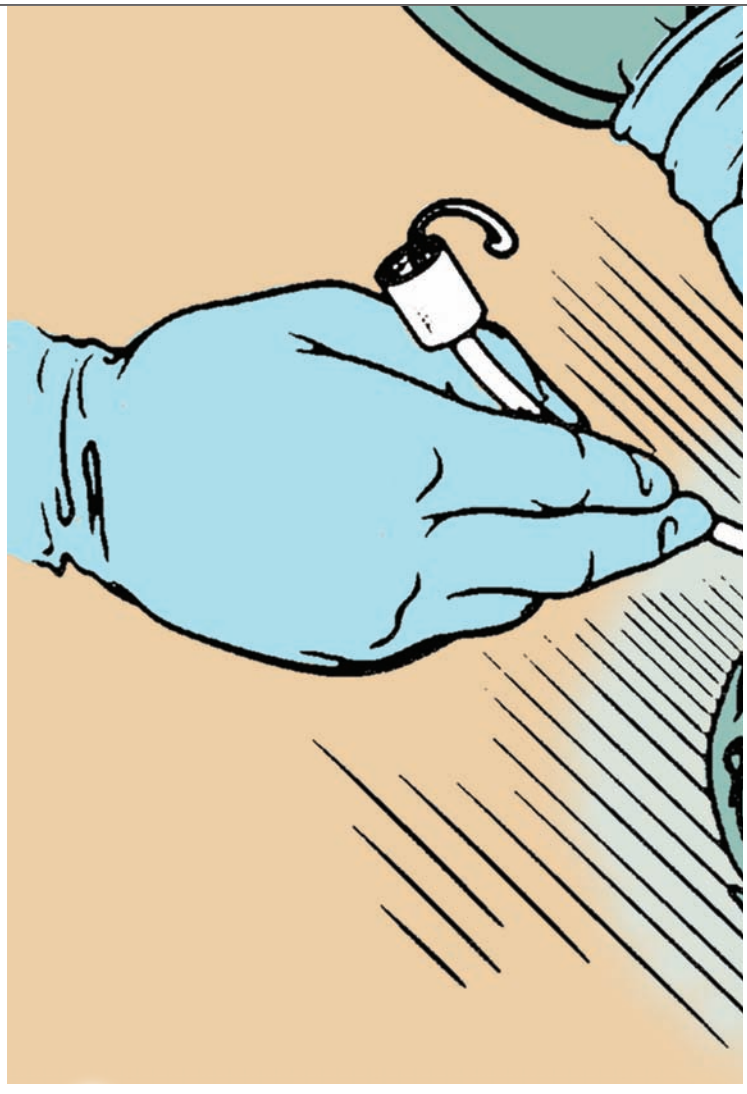

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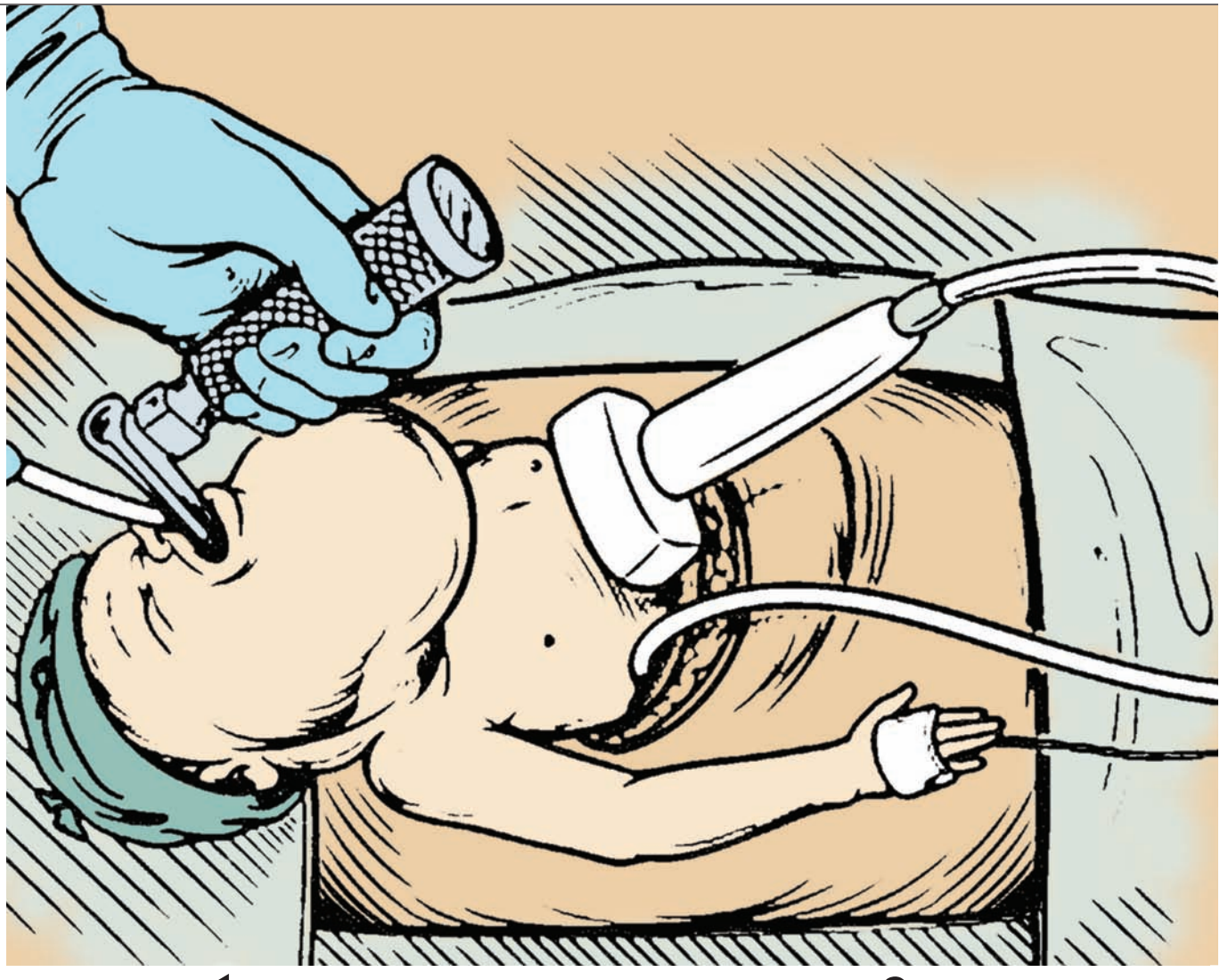
An organized approach to CHAOS and related causes of fetal airway

Learn about your role in delivery of a fetus with life-threatening congenital high airway obstruction syndrome.

By Chuck Biddle, PhD, CRNA, and John Aker, DNAP,CRNA

Congenital high airway obstruction syndrome (CHAOS), originally described by Hedrick and colleagues, is a complete or near-complete airway obstruction identified prenatally via ultrasonography.[1] Between the late 1980s and mid-1990s, case reports began to appear in the literature describing this extremely rare and unvaryingly fatal anomaly. Improvements in fetal imaging now allow early recognition, and the development of the ex utero intrapartum treatment (EXIT) procedure has resulted in



obstruction

the survival of a few infants.[2] Only three fetuses with an antenatal diagnosis of CHAOS are known to have survived following delivery via the EXIT procedure.[3-5]

Types of fetal airway obstruction

Fetal airway obstruction is a life-threatening event with high mortality.[6-8] Difficulty and delay in establishing a patent airway and ventilation is associated with hypoxia, acidosis, and anoxic brain injury leading to life-long disability. Fetal airway obstruction may be intrinsic or extrinsic (see *Causes of fetal airway obstruction*).[9]

CHAOS is a rare presentation of intrinsic obstruction of the larynx or trachea and may be identified prenatally via ultrasonography.[1] The development of the fetal larynx begins during the third week of gestation. Epithelium rapidly proliferates within the laryngotracheal tube and temporarily obliterates the lumen of the larynx. By the

10th week, the larynx recanalizes, leading to laryngeal patency. The failure of this epithelium to recanalize leads to the development of laryngeal atresia (failure to become a tubular structure), laryngeal web, and laryngeal cysts.

This upper airway obstruction causes retention of fetal tracheal fluid, resulting in airway dilation distal to the site of obstruction. The fetal lung produces fluids essential to pulmonary development, including the phospholipid surfactant.[10,11] These pulmonary secretions are normally excreted into the amniotic fluid, but if the fetus has an airway obstruction, the secretions are retained in the lung. This produces a progressive lung expansion that flattens and ultimately inverts the diaphragm.[12] With continued lung expansion, esophageal compression occurs, impairing the fetus' ability to swallow amniotic fluid and leading to polyhydramnios. Fetal ascites and nonimmune hydrops (the abnormal accumulation of fluid in two or more fetal compartments) occur with continued lung expansion impairing venous return and subsequently compressing the heart and vena cava.

Ultrasound findings characteristic of CHAOS and other forms of fetal airway obstruction include high airway obstruction, airway dilation below the obstruction, large echogenic lungs, inverted diaphragm, polyhydramnios, ascites, and nonimmune hydrops. These diagnostic clues may be further clarified with magnetic resonance imaging (MRI), which also can localize the site of airway obstruction.[13] Determining the site of airway obstruction permits precise planning for airway management during the EXIT procedure.[14]

Causes of fetal airway obstruction[12]

Intrinsic—CHAOS
- Laryngeal atresia (the most common etiology of CHAOS)
- Laryngeal cyst or web
- Tracheal atresia
- Subglottic stenosis

Extrinsic
- Cervical teratoma
- Lymphatic malformation
- Vascular anomalies such as vascular rings or a double aortic arch

Cervical teratoma is the most common etiology of extrinsic airway obstruction. Other less common sites for the development of teratoma are the head or neck. These tumors are located anteriorly and comprise cystic and solid tissue that originates from either the thyrocervical area, the palate, or the nasopharynx.[15] As in CHAOS, polyhydramnios is a common finding due to esophageal compression.

Lymphatic malformations, another extrinsic cause of fetal airway obstruction, arise posteriorly, involve the axilla and lateral chest wall, and may have intrathoracic extensions. Between 30% and 70% of fetuses have an accompanying chromosomal abnormality.[12] Lymphatic malformations are typically accompanied by fetal hydrops.

Vascular anomalies may produce external compression of the airway leading to features that are suggestive of CHAOS.[9] Ultrasonography, fetal MRI, and fetal echocardiography can be used to identify the specific extrinsic vascular anomaly. A double aortic arch produces compression of the trachea and esophagus, as the right and left aortic arches encircle the trachea.

While diagnostically different than CHAOS, extrinsic airway obstruction, like CHAOS, may be managed using the EXIT procedure, maintaining uteroplacental circulation until fetal airway patency is established via tracheostomy or endotracheal intubation. With CHAOS, fetal oxygenation and ventilation may require fetal vascular access for extracorporeal membrane oxygenation (ECMO).

Unexpected development

Consider Ms. L, 26, who presented for ultrasound at 20 weeks gestation. She said that she had a previous pregnancy and her baby died at birth. The delivery occurred in a rural hospital and attempts to retrieve medical records were unsuccessful. Ultrasound revealed a fetus with ascites, enlarged lungs, inverted diaphragm, and what appeared to be an abnormally large and obstructed upper airway due to cervical teratoma. By the 32nd week of gestation, the fetus developed hydrops (fetal heart failure); the decision was made to deliver the fetus at 35 weeks gestation using the EXIT procedure. In some cases, open fetal surgery may be considered if the anomaly is diagnosed before 30 weeks.

The operative team consisted of the obstetrician, assistant obstetrician, anesthesiologist, nurse anesthetist, general pediatric surgeon, pediatric

ear-nose-throat (ENT) surgeon, neonatologist, a neonatal ICU (NICU) nurse, an ultrasonographer, two OR circulating nurses, and two scrub persons (see *A typical EXIT team*). One scrub person was dedicated to the maternal procedure team and the second to the fetal procedure team.

Ms. L was brought into the OR, which had been warmed to 86° F (30° C) per facility protocol. She was positioned on the table in a slight left lateral decubitus position to minimize compression of the inferior vena cava by the gravid uterus. Bilateral lower-extremity intermittent pneumatic compression devices were applied. Ms. L was rapidly induced with propofol and a muscle relaxant after breathing 100% oxygen for 10 minutes. She was endotracheally intubated and general anesthesia was administered and titrated to produce profound uterine relaxation. Careful attention to BP, including pharmacologic BP support, ensured uninterrupted and satisfactory placental circulation to the fetus. I.V. fluids were infused through a fluid warmer. No additional tocolytic agents were required but were available in case uterine contractions started.

A Pfannenstiel incision was made and intraoperative ultrasound was used to identify the placental boundaries so that they wouldn't be disturbed by the hysterotomy. Traumatizing the placenta or provoking its detachment jeopardizes placental circulation to the fetus that must be preserved during the EXIT procedure. The hysterotomy was performed and absorbable uterine staples used to ensure hemostasis. The fetal head and upper torso (to about the fetus' nipple line) were delivered into the operative field after careful dissection and opening of the uterus. Scalp electrodes were placed to monitor fetal heart rate and a pulse oximeter was placed on the right upper limb. Umbilical blood flow was assessed by ultrasound.

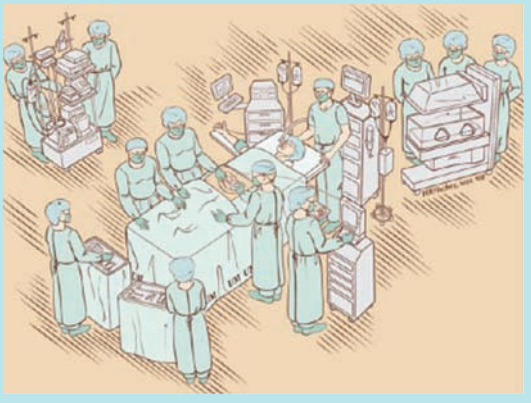
Minimizing fetal exposure reduces fetal heat loss and reduces the risk of premature placental detachment. Laryngoscopy of the fetus using a standard 0 Miller blade proved impossible due to the extremely underdeveloped mandible, cleft palate occupied by a large tongue, and the large cervical teratoma. A pediatric bronchoscope was then used without success as a solid, whitish bulky mass was observed just distal to the vocal cords. At this point, traditional and conservative airway interventions were abandoned and a transverse neck incision was accomplished to create a tracheal stoma. A 2.5 mm (internal diameter) uncuffed endotracheal (ET) tube was placed through the tracheal stoma for surfactant administration, ventilation, and airway pressure monitoring. (The ET tube was eventually replaced by an appropriately sized tracheostomy tube.)

About 6.5 mL of dense mucus and fluid was suctioned via the trachea, which appeared anatomically normal; surfactant was then administered. The circulating nurse provided the pediatric surgeon with a sterile bag-valve–mask device, connected to an external compressed air cylinder with an oxygen source immediately available. Connecting the bag-valve–mask device to the ET tube at the tracheostomy site allowed for gentle hand ventilation of the fetus and confirmation of bilateral breath sounds. Once the tracheostomy site and ET tube were secured, the fetus was fully delivered and the umbilical cord was clamped and severed. The duration of placental support (from head delivery until cord clamping) was 23 minutes. Due to the deep level of maternal anesthesia, which also affected the fetus, Apgar scores weren't assessed, but the fetal heart rate remained between 140 and 150 beats/minute (within normal range) and the fetus' SpO_2 ranged from 88% to 92%, also within the normal range.

Once the neonate and placenta were delivered, the obstetrical team closed the mother's surgical

A typical EXIT team

The operative team typically includes an obstetrician, anesthesiologist, general pediatric surgeon, pediatric ENT surgeon, neonatologist, NICU nurse, ultrasonographer, two OR circulating nurses, and two scrub persons.



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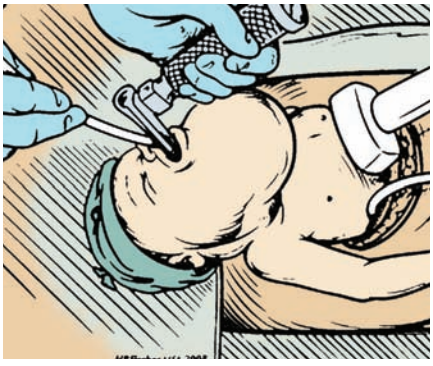
wound, and she recovered uneventfully. The neonate was rapidly transferred to the NICU and placed on pressure-controlled ventilation and positive end-expiratory pressure through the tracheostomy tube that had been placed. An additional dose of surfactant was administered. Ongoing ventilatory support was based on arterial blood gas analysis. Over the subsequent 2 months, the infant was gradually weaned from full, invasive ventilatory support; oral feedings and bilevel positive airway pressure (BiPAP) ventilation were instituted.



The EXIT procedure demands a multidisciplinary team familiar with all aspects of obstetrical surgery.

A collaborative training program involving social workers, NICU nurses, respiratory therapy, pediatric nursing staff, and pediatricians helped the parents learn home care for the infant, who continued to receive BiPAP and had multiple care issues. After gaining a reasonable amount of weight, the infant was discharged from the hospital at 12.5 weeks and the multidisciplinary team (with representatives from pediatrics, ENT, anesthesiology, surgical nursing, and social work) planned for subsequent readmission and surgical airway reconstruction. Home healthcare nursing issues were arranged via the hospital social workers, to provide a bridge from hospital to home.

Unfortunately, 9 days after hospital discharge, the infant, who had been doing well on BiPAP, experienced a sudden respiratory arrest. Despite initial, apparently successful, resuscitation by the parents, subsequent efforts by the arriving emergency medical technicians were unsuccessful. Autopsy revealed severe underdevelopment of the larynx (known at time of delivery) and unexpected diffuse alveolar congestion suggestive of an acute onset of heart failure.

Uses for the EXIT procedure

The EXIT procedure is a highly orchestrated, multidisciplinary team effort. Once candidates are identified in the prenatal period, accurate and detailed diagnosis is managed with high-resolution fetal ultrasound, MRI, and echocardiography. The team approach described in the case report illustrates the need for meticulous advance planning. Once in the OR, the team's full attention is directed at maintaining blood flow to the fetus. Once anesthesia is induced the goal is to achieve uterine relaxation sufficient to prevent contractions and to maintain physiologic uteroplacental circulation, thus ensuring fetal oxygenation. The mother is fully monitored by ECG, cuff and intra-arterial BP, pulse oximetry, and end-tidal CO_2 measurements Neuromuscular function also is monitored.

Although team makeup varies from case to case, the multidisciplinary approach is key. At least two circulating nurses are needed because of the intensive and multitasking nature of the procedure. The scrub persons should be prepared for a variety of anticipated and unexpected interventions. For example, polyhydramnios may require uterine decompression to better assess placental position via ultrasound. Fetal ascites or cystic masses may need to be decompressed if they jeopardize fetal expression from the uterus.

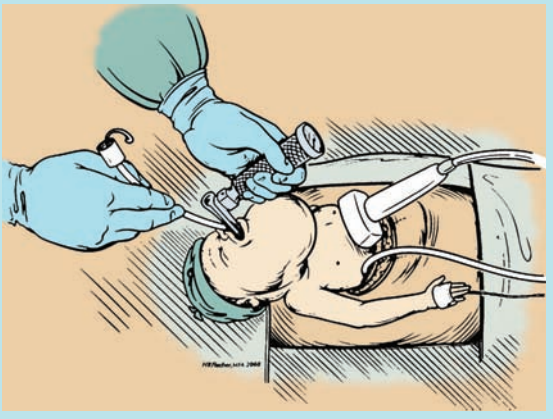
The warm OR temperature helps avoid temperature challenges once the fetal head and partial upper torso are delivered. Because of placental transfer of anesthetic, the fetus is in a state of anesthesia, but additional agents may be administered to the fetus either I.M. or I.V. Once the fetus is delivered, hemoglobin oxygen saturation and heart rate are monitored with pulse oximetry, scalp electrodes, or using continuous echocardiography.

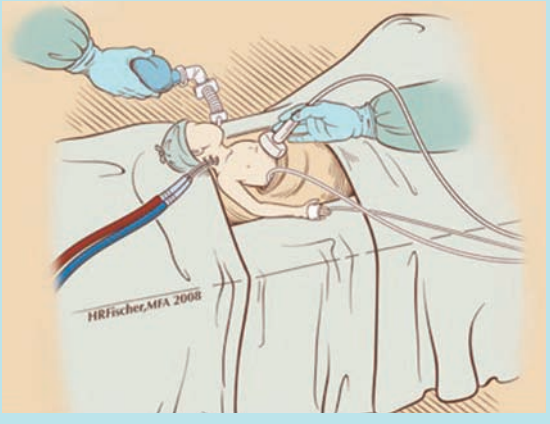
To maintain uterine volume during the procedure, most of the fetus is kept within the uterus and warmed lactated Ringer's solution is instilled into the uterus using either pressure bags or a level 1 fluid infuser. Should fetal distress occur at any time, look for factors that could compromise uteroplacental perfusion, such as umbilical cord compression, inadequate uterine volume, and changes in maternal physiology such as hypotension or hemorrhage. In severe cases I.V. access to the fetus may be needed, so have a sterile setup for rapid fetal I.V. access available.

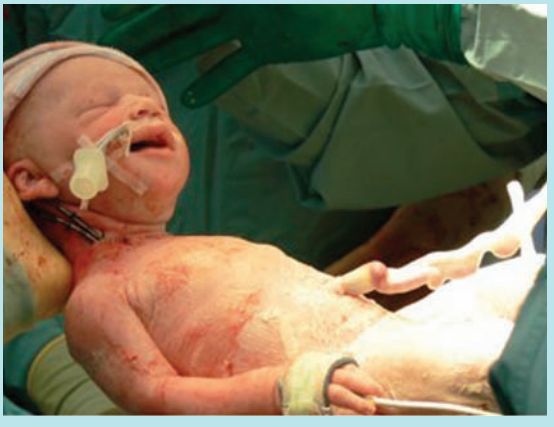
The entire nursing team must be familiar with the anesthetic goals (maternal and fetal issues), the

Monitoring the fetus

During the EXIT procedure, a fetus with CHAOS and other anomalies is monitored with continuous echocardiography (right). ECMO is used when pulmonary or cardiac failure occurs or is anticipated (below). The photograph shows a neonate after a successful EXIT procedure (the infant survived and had a full recovery). Note the cannulation for ECMO (the fetus had significant heart failure) and the pulse oximetry probe on the right hand. Although an ET tube is present, ECMO was needed to avoid barotrauma and permit medical management of heart failure.







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obstetrical surgery goals (delivery of the infant, safety of the mother, maintaining uteroplacental perfusion), and the fetal goals (adequate exposure, monitoring, access to the airway, maintaining physiology). The team should also be familiar with the specialized equipment used, such as ultrasound, uterine stapling devices, and pediatric surgical equipment.

The approach to securing the fetal airway is case and condition specific, and may include basic laryngoscopy with ET intubation, a surgical tracheostomy, or in the worst-case scenario, surgical vascular access for ECMO. Equipment for ECMO should be on standby if the prenatal workup suggests it might be needed (see *Monitoring the fetus*).

Once the fetal airway has been secured and the relative homeostasis of the fetus and mother are assured, the umbilical cord is clamped and the pediatric team manages the neonate. The surgical team is then focused on the surgical closure of the mother as the anesthetic depth is lessened, blood loss is assessed, and uterine tone increased with I.V. oxytocin.

Similar to surgery to separate conjoined twins, which also requires high-level teamwork, the EXIT procedure demands a multidisciplinary team familiar with all aspects of obstetrical surgery, fetal and neonatal surgery, anesthesia care, and physiologic resuscitation. The challenges to the OR nursing staff are formidable in terms of coordination of the team's activity, ensuring availability and use of equipment, maintaining sterility when two mutually exclusive operative procedures are occurring simultaneously, and attending to the additional needs of the surgical and anesthesia staff. Those involved must be capable of a high degree of personal stress management due to the nature of the care provided (and team training should focus more on stress management for team members).

Prepare for the rare

Perioperative nurses play a vital role in the coordinated effort to care for a fetus with CHAOS or another form of fetal airway obstruction. Although the outcome isn't always successful, the EXIT procedure offers hope in selected cases. By understanding these rare fetal anomalies and how they're handled, you can be prepared should they occur. **OR**

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