Ex-Utero Intrapartum Treatment-to-Airway for Obstructing Fetal Neck Masses: A Singular Methodology for Monochorionic and Dichorionic Twin Pregnancies

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Abstract

Fetal airway obstruction in one twin of a diamniotic pregnancy presents unique challenges. Very few cases of ex-utero-intrapartum-treatment (EXIT) procedures for twin pregnancy have been reported and only in dichorionic pregnancies. We report a singular methodology for EXIT-to-airway procedures in two pregnancies involving monochorionic and dichorionic twins.

Two cases of EXIT-to-airway in twin pregnancies were performed in 2018 and 2019 at a regional fetal treatment center. Case 1 involved a giant cervical teratoma in a monochorionic-diamniotic twin pregnancy with preterm labor at 29 weeks. Case 2 involved a dichorionic-diamniotic pregnancy with a large cervical lymphatic malformation with preterm labor at 36 weeks. In each case, the polyhydramnios caused the affected twin's amniotic sac to be the presenting sac for the surgical approach. Bronchoscopy and successful intubation was completed after 22 and 10 minutes of uteroplacental bypass, respectively. The bystander twins were delivered second without intubation and resuscitated without perinatal distress.

EXIT-to-airway appears to be a reasonable option for twins including monochorionic pregnancies, via delivery of the affected twin first followed by delivery of the bystander twin. Thoughtful preparation and counseling by an experienced multidisciplinary team permits an EXIT-to-airway approach for twin pregnancies even in an emergent setting.

Keywords

- EXIT-to-airway
- fetal neck mass
- monochorionic
- dichorionic
- lymphatic malformation
- cervical teratoma
- twin pregnancy
- airway obstruction

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Congenital neck masses can obstruct the fetal airway during development, compromising the ability of the newborn to breathe at birth which carries a high risk for hypoxic brain injury and death.[1,2] The ex-utero intrapartum treatment (EXIT) procedure was developed to prevent catastrophic airway obstruction occurring at the time of birth and has been successfully applied to the treatment of congenital neck masses.[3–6] In contrast with a normal cesarean delivery, an EXIT procedure requires general anesthesia and the administration of tocolytic medications to maintain uterine relaxation and stable uteroplacental circulation.[7] Stable gas exchange through the umbilical circulation provides time to secure the airway before delivering the baby.[8] Fetal airway obstruction in twin pregnancy presents a unique challenge, especially in monochorionic pregnancies, as disruption of the placental circulation during delivery of the affected twin invariably exposes the bystander twin to increased risk. Very few cases of EXIT procedures for twin pregnancy have been reported.[6,9–12] Here, we report a singular methodology for EXIT-to-airway procedures in both monochorionic and dichorionic twin pregnancies.

Case Presentation

Case 1

A 33-year-old G6P2 patient was referred at 26 weeks gestation for the evaluation of her monochorionic-diamniotic pregnancy complicated by a large neck mass in one twin (Twin B). A large, exophytic, mixed solid, and cystic mass was found on fetal ultrasound and magnetic resonance imaging (MRI; ►**Fig. 1**) arising from the anterior neck. There was compression of the cervical trachea and massive polyhydramnios on the side of the affected fetus (deepest vertical pocket [DVP] = 20 cm) and normal DVP on the side of Twin A (4.6 cm). No other anomalies were identified, and there were no insufficient criteria for the diagnosis of twin–twin transfusion syndrome. The bystander twin (Twin A) was normal. The diagnosis of a cervical teratoma was made, and the patient was counseled by the multidisciplinary team about the potential risk of airway compromise of Twin B at birth. Given the size of the neck mass and the need for cesarean delivery, an EXIT-to-airway procedure was recommended at the earliest 28 weeks. Amnioreduction was discussed; however, preference was given toward indomethacin for fetal antidiuretic effect, as the vasa previa present in Twin A increased the risk of complication during amniocentesis. The risks and benefits of the procedure for the mother and both babies were discussed. The massive amniotic sac established Twin B as the presenting twin for any approach. A plan for airway management of Twin B while on placental bypass with dual fetal monitoring was created by the multidisciplinary team including pediatric and fetal surgery, maternal–fetal medicine, neonatology (two teams), obstetrical and fetal anesthesiology, otolaryngology, and pediatric cardiology.

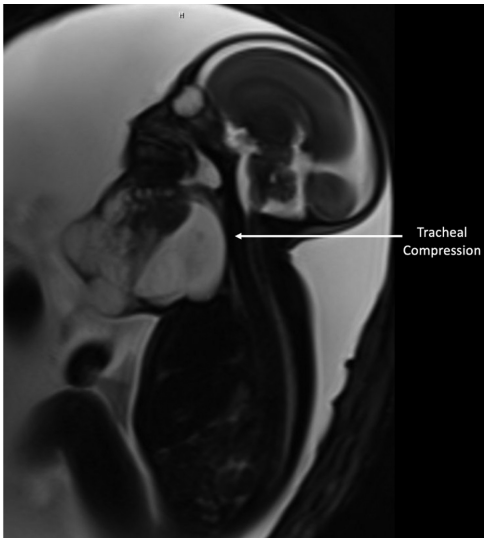

Fig. 1 Fetal MRI at 26 weeks gestation demonstrating a large exophytic, mixed solid and cystic mass in Twin B resulting in tracheal compression.

At 29 weeks gestation, the patient presented with preterm labor. After a brief period of observation, contractions increased in frequency and intensity. Cervical dilation and significant effacement followed. The decision was made to perform an emergent EXIT-to-airway procedure. The entire multidisciplinary team participated in the case. A total intravenous anesthetic (TIVA) was administered and then transitioned to deep-inhaled desflurane prior to the hysterotomy.

Through a midline incision, the uterus was exposed, and the placenta was carefully mapped with ultrasound. An anterior fundal vertical hysterotomy was made to enter the amniotic sac of Twin B. The fetal head and upper torso were delivered (►**Fig. 2A**). An intramuscular cocktail of fentanyl, atropine, and rocuronium was administered to the fetus. Uterine volume was maintained with continuous replacement with lactated Ringer's solution. Continuous monitoring of both fetuses was obtained via transuterine echocardiography (Twin A) or transthoracic echocardiography and pulse-oximetry (Twin B). With anterior displacement of the tumor, rigid bronchoscopy was successfully performed. A 2.5-mm endotracheal tube was passed (►**Fig. 2B**). With hand ventilation, fetal oxygen saturation rose above 90%. The umbilical cord was divided and the neonate was delivered to the neonatology team for resuscitation. Fetal exposure time for Twin B was 22 minutes prior to delivery. Twin A was delivered second, intubated and resuscitated by the neonatology team. The mother and Twin A recovered from the EXIT procedure uneventfully. Following resuscitation, Twin B underwent resection of the large cervical teratoma (►**Fig. 2C,D**) and was discharged after a subsequent tracheostomy and 6-month NICU stay. Twin B eventually required tracheal reconstruction and was later decannulated. Currently, both twins are developing appropriately, as evidenced by the achievement of proper developmental milestones on the Survey of Well-being of Young Children and normal scores on the Preschool Pediatric Symptom Checklist (PPSC) performed at 48 months (PPSC scores < 9, respectively).[13,14] They are both attending preschool and are independent of oxygen or airway support.

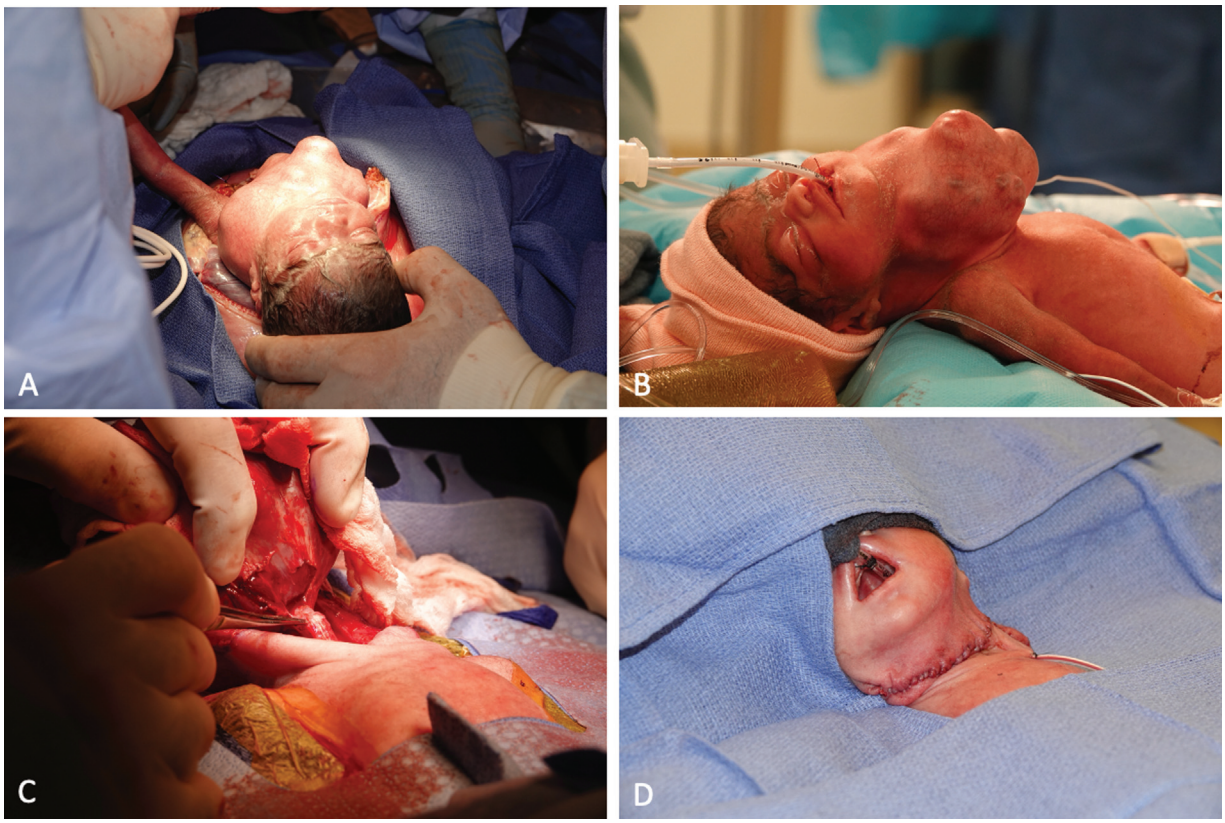

Fig. 2 Intraoperative photos of the EXIT procedure and neck mass resection. (**A**) Twin B partially delivered while remaining on uteroplacental bypass, with a large neck mass resulting in tracheal compression. (**B**) Successful intubation of Twin B. (**C**) Resection of the multicystic mass and dissection from the anterior surface of the trachea. (**D**) Final surgical incision after resection.

Case 2

A 33-year-old G2P1 patient was referred at 24 weeks gestation for the evaluation of her dichorionic-diamniotic pregnancy with one twin (Twin B) affected by a large cervical mass. On fetal ultrasound and MRI, a $10 \times 6.4 \times 5.9$ cm multicystic mass was identified in Twin B involving the anterior and lateral left neck extending to the floor of the mouth and hypopharynx. Polyhydramnios was present (DVP = 10 cm). The remainder of the fetal evaluation was reassuring without any other malformation detected. The diagnosis of multicystic lymphatic malformation was made. The patient was similarly counseled by the multidisciplinary team about the potential risk for airway compromise of Twin B at birth, and an EXIT-to-airway procedure was recommended. The risks and benefits of the procedure for the mother and both babies were discussed. A similar algorithm was developed for the EXIT procedure.

Because of the progressive polyhydramnios and twin gestation, the EXIT-to-airway procedure was scheduled at 36 weeks of gestation. A similar multidisciplinary team attended the procedure, and a similar TIVA/desflurane anesthetic was administered. Through a midline incision, the uterus was exposed, and the placenta was carefully mapped with ultrasound. A posterior fundal hysterotomy was made to selectively enter the amniotic sac of the affected Twin B and expose the fetal head. The fetal head and upper torso were delivered and fetal anesthetic cocktail administered. Continuous monitoring of both fetuses was obtained via transuterine echocardiography (Twin A) or transthoracic echocardiography and pulse-oximetry (Twin B). An endotracheal tube was passed with the assistance of rigid bronchoscopy. With hand-ventilation, the fetal oxygen saturation rose above 90%. The umbilical cord was divided and the neonate delivered to the neonatology team for resuscitation. Fetal exposure time for Twin B was 10 minutes prior to delivery. The bystander twin (Twin A) was delivered second, intubated and resuscitated by the neonatology team. The mother recovered from the EXIT procedure uneventfully. After delivery, Twin A was extubated within 1 hour after admission to the neonatal intensive care unit, rapidly weaned to room air, and was discharged from the hospital without issue after 2 days of observation. Twin B underwent partial resection of the neck mass at 4 months and is currently undergoing sclerotherapy for his multicystic lymphatic malformation. Both children are developing appropriately, with achievement of all developmental milestones on formal evaluation by their pediatrician at 2 years old.

Discussion and Conclusion

In fetal surgery, the pregnant patient is always considered the primary patient, entitled to take the risk for the potential benefit of her fetus. Twin gestation represents a unicum in fetal and perinatal medicine, especially when one of the twins presents a severe congenital malformation. In twin pregnancy affected by a congenital malformation, the health

of the co-twin impacts the decision-making, as the mother and health care providers may accept additional risk for an otherwise healthy fetus.

These cases demonstrate the complex decision-making required for the management of fetal neck masses in twin pregnancy. In one fetus, the lesion would compromise the fetal airway at birth, potentially leading to hypoxia and death. However, treatment of the affected twin via an EXIT procedure introduces the potential for compromise of the bystander twin, which could otherwise be minimized via alternative approaches. The EXIT-to-airway technique allows for adequate airway management and has the potential to prevent respiratory distress in the affected fetus but presents a risk of maternal blood loss, placental abruption, or other acute causes of uteroplacental insufficiency that compromise fetal oxygenation.[15–18] These risks augment with the duration of the procedure.[19] Thus, the EXIT procedure benefits the affected twin, but portends minimal if any advantages for the bystander fetus. On the contrary, the risks of this approach fall on both babies, so the advantages must be weighed not only against the risk for the mother, but also against the additional risk for the bystander twin.

It is important to emphasize that both procedures were performed by a multidisciplinary team with experience in hundreds of cases of open fetal surgery. Detailed preparation and seamless communication are essential elements for safety and success. The decision to proceed is thoroughly vetted among this team and with the patients. When presenting this option to the parents, the health care providers must place a significant effort on education of the mother, and each team must deliver an important perspective of different perioperative risks and benefits. Additional review by an independent ethical oversight committee is essential.

Beside the surgical insult, one must also consider the impact of prematurity. EXIT procedures are scheduled as late as possible in gestation, but concurrent circumstances may force an earlier delivery. For example, the polyhydramnios accompanying obstructing fetal neck masses can prompt preterm labor.[20] Therefore, preterm delivery should be anticipated and the care plan developed accordingly. In order to avoid emergent and unplanned delivery, EXIT is often performed earlier than the due date but realistically, we feel that such a procedure would be appropriate only after 28 weeks gestation. Nevertheless, this intervention can place the burden of prematurity on the healthy bystander twin. If prematurity could be acceptable for a newborn facing a near certain death, the similar risk imposed to a normal fetus raises another serious ethical dilemma.

In the present paper, we demonstrate that EXIT-to-airway procedures are feasible even when twins share a placenta. Additionally, we demonstrate that protection of the bystander twin does not require its delivery first. On the contrary, due to the polyhydramnios often present in this condition, it is likely that the affected twin's amniotic sac will be encountered first during the hysterotomy and complicated maneuvers to avoid it may ultimately compromise control of the procedure. In a prior twin-EXIT case, Liechty et al noted that uterine tone increased after clamping the bystander twin's umbilical cord, requiring additional tocolytic medication and potentially allowing less time to treat the affected twin.[21] Rather, a well-controlled hemostatic entry into the affected twin's sac preserves adequate uteroplacental perfusion for both twins. Simultaneous cardiac monitoring of both fetuses and an alternate rescue algorithm provide measured safety and yield outcomes similar to EXIT procedures in singleton pregnancies.

In conclusion, even in monochorionic-diamniotic gestation, EXIT-to-airway procedures are feasible with good outcomes for the mother and twins. Establishing the airway and delivery of the affected twin prior to delivery of the bystander twin is safe and facilitates stable uteroplacental perfusion during the delivery. We also strongly advise continuous cardiac monitoring of both fetuses throughout the procedure and alternative delivery of both babies with the development of fetal distress. A multidisciplinary approach and advanced planning are paramount for optimal outcomes.

Ethical Approval

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Author Contributions

S.P., F.S., and A.S were responsible for conception and design of study. S.P., F.S., and A.S. performed acquisition of data (laboratory or clinical). F.S. performed data analysis and/or interpretation. F.S., S.P., and A.S. drafted the manuscript. X.F., A.A., K.C., and J.R. provided critical revision. A.S. provided approval of final version of manuscript.

Data Availability

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

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Conflict of Interest

None declared.

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