



# Ex utero intrapartum treatment procedure in two fetuses with airway obstruction

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The *ex utero* intrapartum treatment (EXIT) procedure was introduced to reduce fetal hypoxic damage while establishing an airway in fetuses with upper and lower airway obstruction. Delivery of the fetal head and shoulders while maintaining the uteroplacental circulation offers time to secure the fetal airway. Here, we report two cases of EXIT procedure for fetal airway obstruction, which were successfully managed with extensive preoperative planning by a professional multidisciplinary team.

**Keywords:** Fetal therapy; Laryngeal diseases; Lymphangioma; Prenatal diagnosis

## Introduction

The rapid advancement of imaging technology during the prenatal period has enabled the diagnosis of fetuses with structural anomalies. In particular, prenatal ultrasound and magnetic resonance imaging (MRI) enable the detection of fetal neck masses and the extent of tracheoesophageal obstruction [1]. Fetuses with anomalies that cause airway obstruction usually have poor outcomes because of the delayed time in securing an airway [2]. Hence, many interventions have been introduced to increase the time spent on airway establishment, and the first *ex utero* intrapartum treatment (EXIT) procedure was performed for a large neck mass [3]. Delivery of the fetal head and shoulders while maintaining the uteroplacental circulation offers adequate time to secure the airway and prevent fetal hypoxia [4]. However, because of the rarity of fetal airway obstruction, only few cases of EXIT procedure have been reported in Korea. Here, we report our experiences of EXIT procedure for fetal airway obstruction, which were successfully performed without any complications.

at 20.3 weeks of gestation with a diagnosis of a fetal neck mass. Ultrasonographic findings revealed a 5.3 cm-sized heterogeneous solid mass on the fetal neck, and color Doppler revealed high vascularity. We diagnosed the mass as a cervical lymphangioma or teratoma. Increasing amniotic fluid index in ultrasonography suggested upper airway compression by the mass. Serial ultrasound examination showed an increase in the tumor size to up to almost 8 cm at 30.3 weeks of gestation (Fig. 1A). To evaluate airway patency, MRI was performed at 31.0 weeks of gestation and revealed an 8.5 cm-sized mass arising from the mandible and anterior neck soft tissues and compressing the upper airway (Fig. 1B). Delivery was performed via a cesarean section with the EXIT procedure at 38.3 weeks of gestation. A multidisciplinary

Received: 2017.05.28. Revised: 2017.07.27. Accepted: 2017.08.07.

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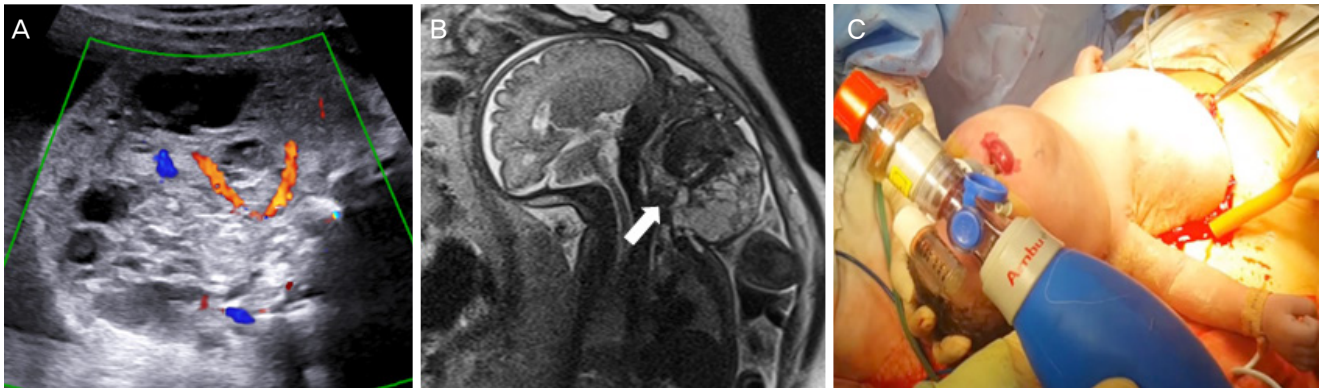
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## Case report

### 1. Case 1

A 32-year-old nulligravida was referred to our institution



**Fig. 1.** Ultrasonographic findings at 30.3 weeks of gestation showing a huge heterogeneous solid mass on the fetal neck with high vascularity (A). Magnetic resonance imaging at 31.0 weeks of gestation showing an 8.5 cm-sized neck mass with suspected airway obstruction (arrow in B). After partially delivering the fetal head and shoulders, intubation was performed (C).

team comprising obstetricians, neonatologists, anesthesiologists, and otorhinolaryngologists was formed. The fetus was in cephalic presentation, and the placenta was located at the posterior body of the uterus. A Pfannenstiel skin incision and a lower uterine segment transverse hysterotomy were performed. The fetal head and upper body were delivered, while the lower body remained in the uterus (Fig. 1C). Compression of the fundus was avoided, and warm saline was continuously infused into the uterus to maintain the intrauterine volume and to prevent placental separation. Peripheral oxygen saturation (SpO<sub>2</sub>) and fetal heart rate were monitored. Initial fetal SpO<sub>2</sub> was 23%, with a heart rate of 100 beats/min. During several intubation attempts, SpO<sub>2</sub> varied from 48% to 56%, with a heart rate of 136 beats/min. Intubation was successfully accomplished in 12 minutes. The baby was then delivered, and the umbilical cord was clamped. The birth weight was 3,500 g, including the neck mass, and the Apgar score was 6 and 7 at 1 and 5 minutes, respectively.

After the delivery of the baby and the placenta, carbetocin was injected, and the uterine contraction was good. Postoperatively, the estimated blood loss was 1.0 L, which was similar to the volume loss during routine cesarean section. The hemoglobin level was 11.8 g/dL preoperatively and decreased to 9.3 g/dL on postoperative day 1. The mother was discharged on postoperative day 3 without any complications.

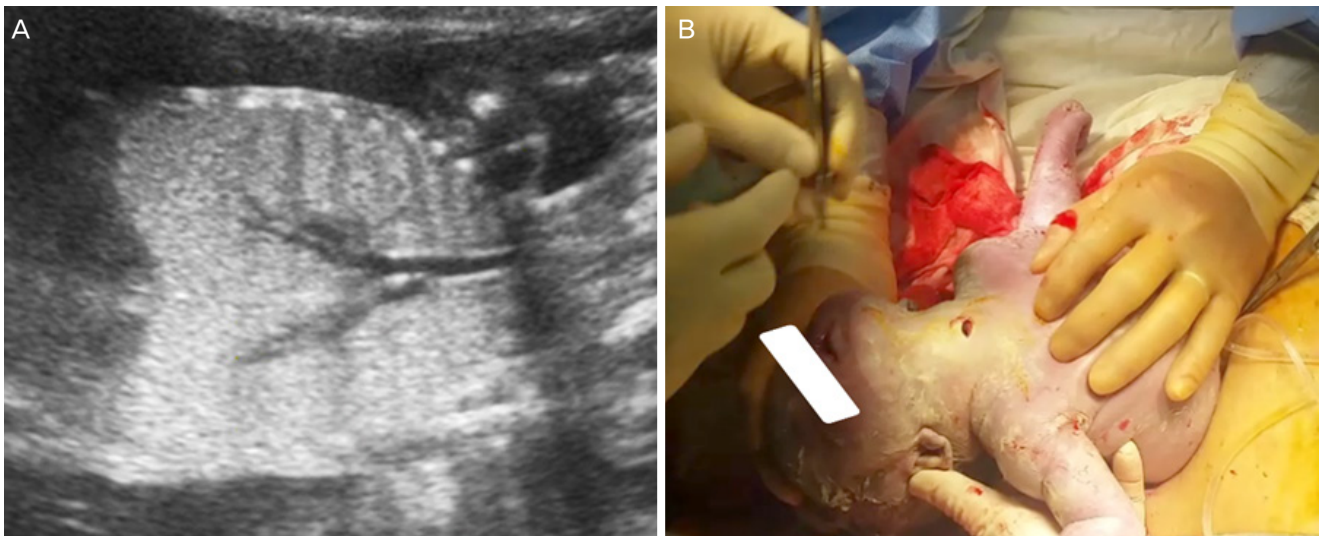
Postnatal MRI demonstrated a large cervicofacial lymphangioma with hemorrhage. Excision of the mass with tracheostomy was performed at 62 days after birth. Complete resection was impossible, and histopathological examination confirmed a cavernous lymphangioma. After repeating several courses of ethanol sclerotherapy, the baby was discharged in toler-

able condition with tracheostomy at 4 weeks postoperatively. Currently, at the age of 2 years, the baby remains in tracheostomy state but is doing well.

## 2. Case 2

A 31-year-old nulligravida was referred at 21.1 weeks of gestation with a suspected diagnosis of congenital high airway obstruction syndrome (CHAOS). Ultrasonography revealed symmetrically enlarged, hyperechoic, and homogeneous lungs with an inverted diaphragm, anteriorly displaced heart, and associated polyhydramnios, all of which suggested CHAOS (Fig. 2A). Delivery was performed via a cesarean section with EXIT procedure at 37.0 weeks of gestation. The fetus was in cephalic position, and the placenta was located in the posterior body of the uterus. A similar operation to that in the case 1 was performed. Initial fetal SpO<sub>2</sub> was 75%, with a heart rate 145 beats/min. Intubation was failed several times because of the invisible airway. The total amount of time spent on intubation attempts was approximately 5 minutes. Tracheostomy was then performed within 3 minutes (Fig. 2B), and SpO<sub>2</sub> recovered to 82%. After approximately 10 minutes, the umbilical cord was clamped. The birth weight was 2,720 g, with an Apgar score of 6 and 8 at 1 and 5 minutes, respectively.

After delivery, the uterus showed poor contraction initially. After injecting carbetocin with a continuous infusion of sulprostone, uterine contraction was normalized; however, the expected blood loss was >2 L. The initial hemoglobin level was 13.2 g/dL, which decreased to 9.6 g/dL on the postoperative day 1. The mother had no postoperative complications and was discharged on postoperative day 3.



**Fig. 2.** Ultrasonographic findings showing both lung hyperexpansion with diaphragm inversion, suggestive of congenital high airway obstruction syndrome (A). Tracheostomy is in process while the baby is partially delivered (B).

Postnatal computerized tomography revealed a short segmental obliteration of the subglottis, with the visible lumen of the proximal trachea, suggesting laryngeal atresia. After conservative treatment during 1 month, the baby was discharged. At the age of 1 year, the baby remains in tracheostomy state but is doing well.

## Discussion

To the best of our knowledge, to date, only five cases involving the EXIT procedure have been reported in Korea [5-9]. Previous reports mainly focused on the anesthetic management of the EXIT procedure to achieve adequate uterine relaxation, maintain uteroplacental circulation, and minimize fetal movement. Furthermore, management after the EXIT procedure has been discussed by pediatricians and otorhinolaryngologists, with a focus on postnatal outcomes. On the basis of our successful experience with the EXIT procedure, we would like to mainly discuss the role of the obstetrician performing the EXIT procedure.

The EXIT procedure aims to manage fetuses with life-threatening diseases of the upper respiratory system, thereby increasing the survival rate of fetuses. Selecting appropriate candidates for the EXIT procedure is primarily important. Indications for the EXIT procedure to secure the airway are as follows: 1) masses that cause extrinsic airway compression such

as teratoma, lymphangioma, and epignathus; 2) severe micrognathia or agnathia; 3) CHAOS; and 4) removal of balloon occlusion in congenital diaphragmatic hernia [10]. The EXIT procedure can also be performed for resecting huge chest masses that cause airway obstruction, such as in congenital pulmonary airway malformation, bronchopulmonary sequestration, and mediastinal or pericardial teratoma [11]. Other indications include severe congenital heart disease or severe congenital diaphragmatic hernia for applying extracorporeal membrane oxygenation and conjoined twins for separation [11]. Obstetricians should be aware of the abovementioned indications. Our second case of CHAOS safely managed with EXIT procedure was first reported in Korea, and our experiences may encourage the parents of fetuses with CHAOS to continue the pregnancy and to receive proper postnatal management.

Once a diagnose is made, the obstetricians should obtain the data of mass size and its location as well as the characteristics of diseases causing airway obstruction. Although ultrasound is a standard tool for determining the need for the EXIT procedure, it has a limitation regarding the evaluation of airway obstruction. MRI is superior to ultrasound for assessing the extent of airway distortion [10]. MRI was not performed in our second case where the baby had only a pinpoint trachea. If MRI was prenatally performed, the time spent on intubation could have been reduced.

The organization of a multidisciplinary team comprising obstetricians, neonatologists, anesthesiologists, and otorhino-

laryngologists is essential for achieving successful outcomes of the EXIT procedure. Before performing the EXIT procedure, our multidisciplinary team gathered, discussed the need for tracheostomy, and simulated the condition during the EXIT procedure. The obstetricians should decide an appropriate time for delivery with considering both fetal and maternal conditions. The EXIT procedure should be performed before the onset of labor. Accordingly, 37–38 weeks of gestation is generally acceptable unless there are any other maternal or fetal indications for earlier delivery [4]. In addition, evaluation about the presence of polyhydramnios, fetal position, and placental location before the EXIT procedure is important. Therefore, obstetricians require careful mapping of the placental edges, which could alter the location of the uterine incision; they also need to determine the degree of exposure required, according to the fetal abnormalities [4].

During the EXIT procedure, obstetricians should focus on maximizing uterine relaxation time by not compressing the uterine fundus, delivering only the head and shoulders onto the surgical field, and infusing warm saline into the uterus to maintain the uterine volume and prevent placental separation and umbilical cord compression [4]. However, excess time spent on establishing the fetal airway during the procedure can increase maternal complications, particularly postpartum hemorrhage. Although uterine atony resulted in an increased estimated blood loss in our second case, it was successfully managed with uterotonics. Because other complications such as uterine rupture or dehiscence at subsequent pregnancy may also occur, an inter-pregnancy interval of at least 24 months following EXIT procedure has been recommended [12].

The EXIT procedure can be a potent procedure for cases of life-threatening airway obstruction. Obstetricians play an important role in managing the EXIT procedure for both fetuses and mothers. Minimizing fetal and maternal complications should be the goal for obstetricians performing the EXIT procedure.

## Conflict of interest

No potential conflict of interest relevant to this article was reported.

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