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ORIGINAL ARTICLE

Clinical outcomes of ex utero intrapartum treatment for fetal airway obstruction

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Abstract

Purpose The aim of this study was to evaluate the indications and the clinical outcomes of the fetuses managed with ex utero intrapartum treatment (EXIT) procedures.

Methods We retrospectively reviewed the medical records of all fetuses who underwent EXIT procedures between 2003 and 2018.

Results EXIT procedures were performed in nine cases. The prenatal diagnosis of the neonates was congenital high airway obstruction syndrome in four cases, the neck masse in five cases. Although the airway management under the EXIT procedure was successful in eight cases, the airway management failed in one case. During the EXIT procedures, the airway was managed by endotracheal intubation in two cases, whereas six cases underwent tracheostomy. Six cases with fetal airway obstruction survived to discharge, whereas three cases died due to airway management failure or complications of the underlying disease. A case with a cervical teratoma underwent tumor resection the day after birth due to rapid enlargement of the neck mass. Long-term survival was achieved in five cases.

Conclusions We concluded that the EXIT procedure was effective and could be performed safely in the airway management of fetuses with suspected airway obstruction. The treatment strategy for the neck masses should be planned before birth.

Keywords Ex utero intrapartum treatment · Congenital high airway obstruction syndrome · Epignathus · Cervical teratoma · Cervical lymphatic malformation

Introduction

Recent advances in prenatal diagnostic imaging, including fetal ultrasonography and fetal magnetic resonance imaging (MRI) have led to changes in perinatal management and improved neonatal outcomes [1]. Fetuses with airway obstruction, such as congenital high airway obstruction syndrome (CHAOS) and giant neck masses arising around the upper airway are at high risk for hypoxic brain injury and death at birth [1–4]. The ex utero intrapartum treatment (EXIT) procedure is a technique that can provide time to manage the fetal airway by intubation or tracheostomy while maintaining placental blood circulation to the fetus [4–9]. The EXIT procedure was initially advocated for the reversal of tracheal occlusion in fetuses with severe congenital diaphragmatic hernias [10, 11]. Recently, management of the fetal airway in the presence of an obstructing mass or in the setting of CHAOS has become a common indication for the EXIT procedure, and the indication has expanded to other

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diseases, such as congenital lung lesions. However, because of the rarity of fetal airway obstruction, the application of the EXIT procedure has only been reported in a few cases in Japan. This study aimed to evaluate the indications and clinical outcomes of fetal airway obstruction, such as CHAOS and giant neck masses arising in the upper airway of patients treated with the EXIT procedure.

Methods

Study design

This study was approved by the institutional review board of Osaka Women's and Children's Hospital (no. 1019) and Osaka University Hospital (no. 18228). We retrospectively reviewed our consecutive experience with the EXIT procedure in the airway management of nine fetuses with fetal airway obstruction such as CHAOS and giant neck masses arising around the upper airway in cases that were managed at Osaka Women's and Children's Hospital and Osaka University Hospital between January 2003 and January 2018. The maternal and infant's medical records were retrospectively reviewed to obtain the demographic information of the patients. The maternal data included the maternal age, gestational age at the initial diagnosis, the polyhydramnios status, patency of the fetal airway, tumor size, findings of fetal imaging, time on placental support, total operative time, hemoglobin decrease, maternal complications, and length of hospital stay. The neonate data included gestational age at birth, sex, birth weight, Apgar score, method of airway management, umbilical cord blood gas, congenital associated anomalies, tumor pathology, length of hospital stay, and the postnatal course and outcomes. The data are presented as the median (range). The patency of the fetal airway was verified by confirming the lumen of upper airway on fetal MRI or the fluid flow associated with fetal breathing on fetal ultrasonography.

Ex utero intrapartum treatment

After the fetal diagnosis was made based on prenatal ultrasonography and magnetic resonance imaging (MRI), obstetricians and pediatric surgeons explained the planned fetal treatments and the expected fetal prognosis to the parents of the fetus. Informed consent was obtained from the parents. We held a multidisciplinary joint conference about the indications for the EXIT procedure, and discussed the optimal methods of airway management. The EXIT procedure was performed by a multidisciplinary team consisting of obstetricians, anesthesiologists, pediatric surgeons, neonatologists, otolaryngologists, oral and maxillofacial surgeons and operating room nurses. The gestational ages at which the EXIT procedure was performed depended on the individual situation. If the mother and fetus were in a stable condition, we planned to perform the EXIT procedure at full term. Most patients undergo placement of a lumbar epidural catheter for postoperative pain relief. The EXIT procedure was performed under general anesthesia to achieve adequate uterine relaxation. Anesthesia is induced by a rapid sequence technique with thiamylal sodium, rocuronium and fentanyl administered given intravenously, followed by endotracheal intubation. High-dose (2–3 minimum alveolar concentration) inhalation agents were used to maintain uterine relaxation. Ephedrine was administered intravenously for maternal hypotension secondary to deep general anesthesia. Once the uterus was exposed, intraoperative ultrasonography was used to confirm the position of the fetus and the edge of the placenta. After lower transverse uterine incision, the sutures were placed at the edge of the uterine incision to prevent massive bleeding. A uterine stapler was not used, since this device is not available in Japan. The uterine cavity was filled continuously with warm physiological saline solution using a rapid infusion pump to maintain an adequate uterine volume for avoiding placental abruption and to prevent compression of the umbilical cord. The fetal head and the upper limb of one side were exteriorized, while the rest of the body remained within the uterine cavity. The fetus was continuously monitored by a pulse oximeter that was placed on the upper limb of the fetus and a sterile probe was placed on the chest of the fetus for intraoperative echocardiography. Fetal anesthesia was administered by the transplacental transfer of maternally inhaled anesthetics and supplemented with the intramuscular injection of vecuronium (0.2 mg/kg), fentanyl (10 μg/kg) and atropine (20 μg/kg) during the EXIT procedure. Once the fetal head and neck were exposed, the airway was identified to perform endotracheal intubation or tracheostomy. When endotracheal intubation failed, tracheostomy was performed. Once the airway was accessed, manual ventilation was initiated during placental blood circulation. After confirmation of carbon dioxide elimination by an end-tidal carbon dioxide detector, the umbilical cord was clamped and divided, then the neonate was transferred to the adjacent room for further resuscitation and evaluation. The uterus and maternal abdomen were closed in a similar manner to cesarean section.

Results

Characteristics of the mothers

A total of nine cases were delivered by EXIT procedures during the 15-year period (Table 1). The median and range of maternal age was 33 (26–38) years. A chromosomal analysis indicated that the karyotypes were normal in five

Table 1 Characteristics of the mothers

Case	Age (year)	Gravidity and parity	Karyotype	GA at initial diagnosis (week)	Prenatal diagnosis	Polyhydramnios (amnioreduction)	Patency of the fetal airway	Findings of fetal imaging
1	38	G2 P1	Normal	22	CHAOS	Yes (done)	No	Bilateral enlarged echogenic lungs, dilated airway, flattened diaphragms, fetal ascites and hydrops
2	28	G2 P1	Normal	20	CHAOS	No	No	Bilateral enlarged echogenic lungs, dilated airway, flattened diaphragms, fetal ascites and hydrops
3	27	G1 P0	No examination	24	CHAOS	Yes	No	Bilateral enlarged echogenic lungs, dilated airway, inverted diaphragms, fetal ascites and hydrops
4	35	G1 P0	Normal	21	CHAOS	Yes	No	Bilateral enlarged echogenic lungs, dilated airway, flattened diaphragms, fetal ascites and hydrops
5	30	G1 P0	Normal	23	Epignathus	Yes	Yes	8.0 × 8.0 × 0.5 cm cystic and solid oropharyngeal mass
6	34	G4 P3	No examination	16	Epignathus	Yes (done)	Yes	14.0 × 15.0 × 6.0 cm cystic and solid oropharyngeal mass with hydrops
7	26	G1 P0	No examination	27	Cervical teratoma	Yes (done)	No	10.0 × 9.0 × 6.0 cm cystic and solid cervical mass with airway compression and distortion
8	33	G2 P1	Normal	26	Cervical teratoma	Yes (done)	Yes	19.0 × 12.5 × 10.0 cm right cystic and solid cervical mass with airway compression and distortion
9	35	G2 P1	No examination	21	Cervical lymphatic malformation	No	Yes	13.0 × 7.5 × 6.0 cm multiple cystic mass extending to upper mediastinum with airway compression and distortion

GA gestational age, *CHAOS* congenital high airway obstruction syndrome

cases. The initial prenatal diagnosis was made at a median and a range of gestational age of 22 (16–27) weeks.All cases were diagnosed by fetal ultrasonography and fetal MRI, and the patency or deviation of the airway was evaluated. Four of the fetuses were diagnosed with CHAOS. Five of the fetuses were diagnosed with a giant neck mass arising around the upper airway, which consisted of epignathus in two cases, cervical teratoma in two cases and cervical lymphatic malformation in one case. The patency of the airway was confirmed in four cases in which a giant neck mass was diagnosed. Seven cases developed polyhydramnios, and four cases among them required amnioreduction (Table 1).

The outcomes of the mothers

EXIT procedures were performed electively in seven cases, and emergently in two cases due to a fetal heart rate abnormality with intrauterine growth retardation at

28 weeks of gestation in case 1, and premature rupture of membranes associated with polyhydramnios at 30 weeks of gestation in case 5 (Table 2). The median and range of gestational age at the time of the EXIT procedure was 34 (28–37) weeks. For the mothers, the median and range of operative time was 74 (44–106) min and the median and range of difference in the hemoglobin level between before and after the EXIT procedure was 2.9 (1.4–4.7) g/dl. No mothers showed hemodynamic instability during the EXIT procedure. No cases required blood transfusion. Two mothers developed surgical wound infection as a postoperative complication, and the median and range of length of hospital stay after the EXIT procedure was 7 (4–30) day, which is the average length of hospital stay after cesarean section in Japan. One mother required prolonged hospitalization after surgery due to wound infection.

Summary of the fetuses treated with EXIT procedures

The median and range of gestational age at delivery of patients treated with EXIT procedures was 34 weeks (range 28–37) (Table 3). The median and range of duration of uteroplacental support, which was defined as the time from uterine incision to umbilical cord clamping, was 18 (8–44) min. Among the nine cases, only one fetus developed hemodynamic instability during the EXIT procedure. We interrupted the EXIT procedure at 28 min in case 7, which involved a patient with giant cervical teratoma due to a fetal heart rate abnormality that was possibly caused by umbilical cord compression. The airway was successfully accessed and managed under the EXIT procedure in the other eight cases. Two cases were finally endotracheally intubated using a laryngoscope with difficulty, whereas five cases required tracheostomy. Although we tried to intubate case 7 after delivery, endotracheal intubation failed due to

Table 2 The outcomes of the mothers

Case	Timing of EXIT	GA at EXIT (week)	Total operative time (min)	Total anesthesia time (min)	Difference in Hb (g/dl)	Blood transfusion	Postoperative complications	LOS after EXIT (day)
1	Emergent	30	57	100	12.6→9.7	None	None	16
2	Electively	37	74	132	10.7→7.3	None	Wound infection	7
3	Electively	37	44	86	12.3→9.7	None	None	7
4	Electively	34	106	169	12.9→8.2	None	None	8
5	Emergent	33	47	108	11.4→8.1	None	Wound infection	30
6	Electively	28	74	113	11.1→7.0	None	None	6
7	Electively	33	101	201	8.9→7.5	None	None	4
8	Electively	36	47	82	10.1→7.7	None	None	6
9	Electively	36	80	127	10.8→7.9	None	None	6

GA gestational age, *EXIT* ex utero intrapartum treatment, *Hb* hemoglobin, *LOS* length of hospital stay

Table 3 Summary of the fetuses treated with EXIT procedures

Case	GA at delivery (week)	Duration of uteroplacental support (min)	Stability of fetal hemodynamics	Airway managements	Umbilical arterial cord blood gas			
					pH	O_2 (mmHg)	CO_2 (mmHg)	BE (mmol/l)
1	30	14	Stable	Tracheostomy	7.286	30.5	48.8	−3.3
2	37	44	Stable	Tracheostomy	7.121	21.5	78.1	−8.1
3	37	12	Stable	Tracheostomy	7.248	27.4	57.8	−3.8
4	34	8	Stable	Tracheostomy	7.154	31.2	71.7	−3.6
5	33	18	Stable	Tracheostomy	7.271	35.9	49.3	−5.2
6	28	22	Stable	Tracheostomy	7.039	9.4	79	−12
7	33	28	Bradycardia	Not achieved	6.956	9	87	−15.5
8	36	17	Stable	Endotracheal intubation	7.119	20.7	68	−9.1
9	36	41	Stable	Endotracheal intubation	7.239	28.6	64.8	−2.6

GA gestational age, *EXIT* ex utero intrapartum treatment, *BE* base excess

the broad expanse of the tumor (from the anterior neck to the oral cavity).

The characteristics and outcomes of the fetuses

The postnatal diagnosis was laryngeal atresia in four cases, epignathus in two cases, cervical teratoma in two cases and cervical lymphatic malformation in one case (Table 4). The median and range of birth weight was 2178 (1064–3512) g. Multiple congenital anomalies were confirmed in two cases of laryngeal atresia; in both cases, the patient died in infancy due to complications associated with respiratory anomalies. All patients with laryngeal atresia require a tracheostomy after discharge from hospital. In case 5, a patient with epignathus underwent tracheostomy under the EXIT procedure and underwent tumor debulking immediately after birth (Fig. 1). The pathology of the tumor was mature teratoma. Resection of the remaining tumor in the oral cavity was performed at 20 and 49 days of age, and the patient was freed from tracheostomy at 46 months of age. He can normally eat food orally at 13 years of age. In case 8, a patient with a giant cervical teratoma underwent endotracheal intubation under the EXIT procedure and underwent tumor resection the day after birth due to rapid enlargement of the neck mass (Fig. 2). A pathological examination revealed immature teratoma. Unilateral vocal cord paralysis and facial nerve paralysis occurred as postoperative complications. He was discharged from our hospital at 3 months of age. In case 9, a patient with cervical lymphatic malformation underwent endotracheal intubation under the EXIT procedure and underwent tracheostomy with partial tumor resection on the day after birth (Fig. 3). The patient underwent sclerotherapy and resection of the anterior mediastinal tumor at 9 months of age due to respiratory failure that was affected by the enlargement of the tumor. He still has microcystic lesion involving the floor of the mouth and the deep space of the neck, which necessitated gastrostomy. At 20 months of age, he can eat a certain amount of food and has been weaned from mechanical ventilator support. In case 6, the patient died of severe bloody effusion from the tumor immediately after birth. In case 7, the patient died immediately after birth due to airway management failure. During the follow-up period, the overall mortality rate was 44.4%.

Discussion

The EXIT procedure is a technique that can provide time to manage the fetal airway while maintaining placental blood circulation to the fetus. During the EXIT procedure, the mother requires general anesthesia and is in danger of increased blood loss due to uterine relaxation, which is

Table 4 The characteristics and outcomes of the fetuses

Case	Postnatal diagnosis	Sex	Birth weight (g)	Apgar score (1 min/5 min)	Associated anomalies	Pathology	LOS (day)	Follow-up period (month)	Outcome
1	Laryngeal atresia	Female	1536	2/6	Congenital bronchial stenosis, duodenal atresia, anorectal malformation	–	258	12	Death
2	Laryngeal atresia	Female	1868	2/2	Cornelia de lange syndrome tracheomalacia, atrial septal defect	–	240	25	Death
3	Laryngeal atresia	Male	3002	2/3	None	–	133	12	Alive
4	Laryngeal atresia	Female	1908	2/5	None	–	130	68	Alive
5	Epignathus	Male	2178	6/6	Cleft palate	Mature teratoma	255	167	Alive
6	Epignathus	Female	1064	4/5	Cleft palate	Immature teratoma	0	0	Death
7	Cervical teratoma	Male	2458	1/1	None	Immature teratoma	0	0	Death
8	Cervical teratoma	Male	3512	3/3	None	Immature teratoma	101	8	Alive
9	Cervical lymphatic malformation	Male	3270	2/3	None	Lymphatic malformation	220	21	Alive

BW birth weight, *LOS* length of hospital stay

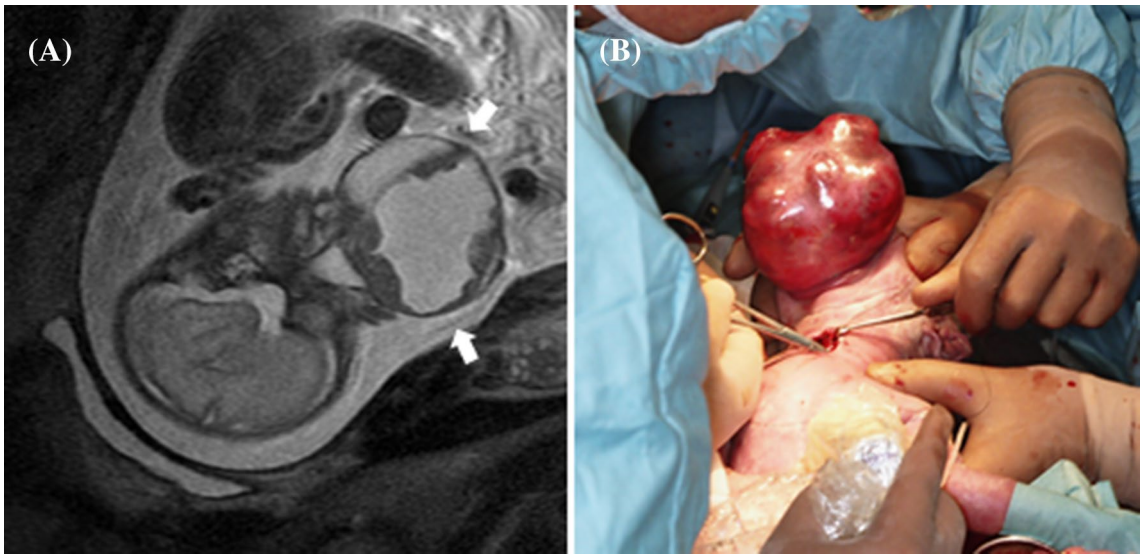

Fig. 1 A case of epignathus (case 5). **a** T2-weighted sagittal fetal MRI at 23 weeks of gestation showed a large solid and cystic oropharyngeal mass originating in the pharynx and protruding outside the mouth (arrow). **b** A fetus undergoing tracheostomy during the EXIT procedure

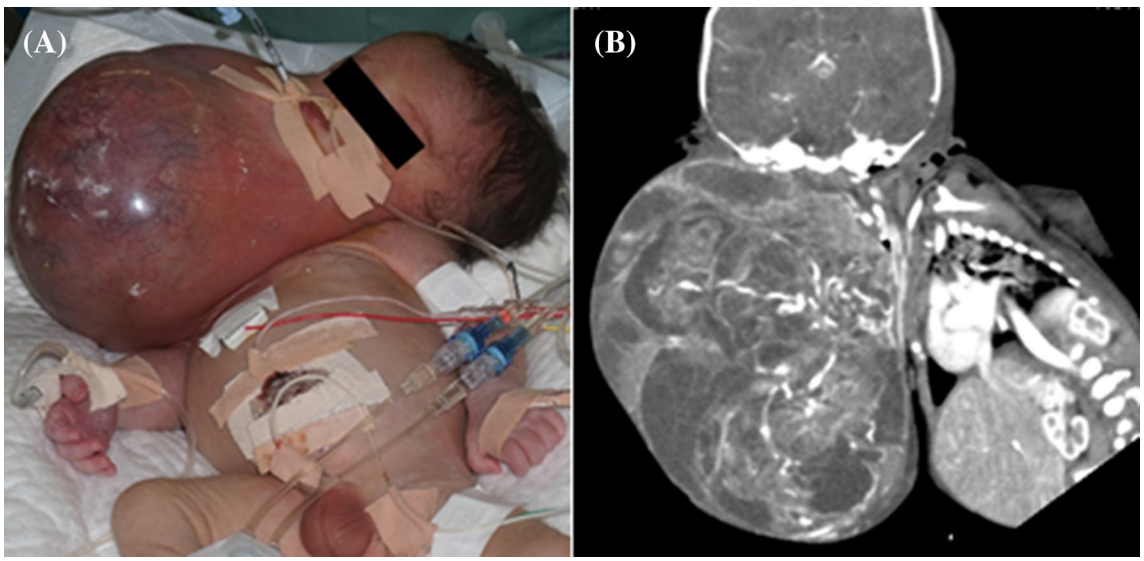

Fig. 2 A case of giant cervical teratoma (case 8). **a** A preoperative view after endotracheal intubation under the EXIT procedure showing the giant neck mass. **b** Computed tomography showed that the giant neck mass had mixed components consisting of a solid part and a cystic part

required to preserve the uteroplacental gas exchange [12, 13]. Placental abruption or placental laceration during the procedure could lead to maternal and fetal hemorrhage [1]. Maternal hemodynamic stability and good oxygenation are essential for maintaining uteroplacental circulation [10]. Thus, the maternal risk becomes higher if the EXIT procedures become longer [6, 14]. In our series, the maternal hemodynamics remained stable for up to 44 min without any significant complications. The median difference in the hemoglobin level between before and after the EXIT procedure was 2.9 g/dl; no cases required blood transfusion. Noah et al. advocated patients who underwent the EXIT procedure had a higher incidence of wound complications in comparison to those who underwent a common cesarean section procedure [14]. Two cases also developed wound complications after surgery in our series. We hypothesize the use of a larger incision and the longer time is required in the EXIT procedure in comparison to standard cesarean section is the reason for this complication. Although a larger uterine incision may increase the risk of uterine rupture during subsequent pregnancies, three mothers who desired further pregnancy have subsequently had uncomplicated deliveries.

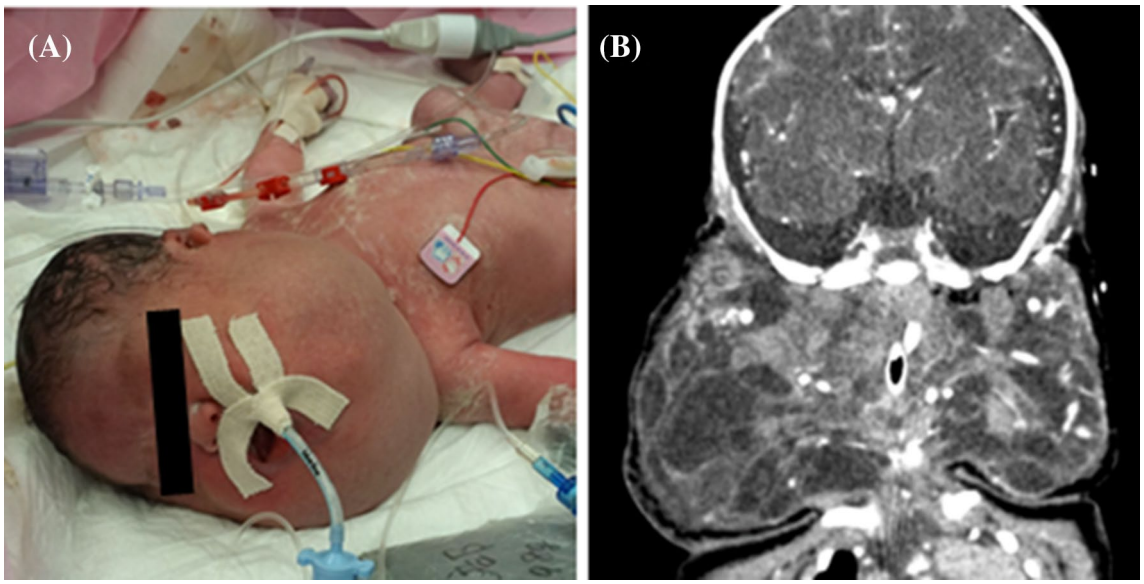

Fig. 3 A case of cervical lymphatic malformation (case 9). **a** The preoperative view after endotracheal intubation under the EXIT procedure in a patient with cervical lymphatic malformation. **b** Computed tomography showed that cervical lymphatic malformation had mixed components consisting of a solid part and a cystic part

The fetal hemodynamics during the EXIT procedure were stable, except for patient with a giant cervical teratoma, who showed a bradycardia during the EXIT procedure. As the fetal heart rate during the EXIT procedure is a good indicator of placental perfusion, we interrupted the procedure on the assumption that the placental perfusion deteriorated. Because the fetal upper body was exteriorized and the umbilical cord was prolapsed from the uterus, umbilical cord compression might have occurred, causing fetal bradycardia. Although an adequate uterine volume was maintained by continuous physiological saline solution filling, we should have been very careful not to be exteriorize the upper body of the fetus from the uterus.

The general indications for the EXIT procedure to secure the airway were as follow: (1) removal of balloon occlusion in severe congenital diaphragmatic hernias; (2) neck and oral masses that causes extrinsic airway compression such as teratoma, epignathus and lymphatic malformation; (3) CHAOS; (4) severe micrognathia. The criteria for applying the EXIT procedure in deliveries in our institution were as follows: evidence of CHAOS deviation/compression/obstruction of the airway, involvement of the floor of the mouth and the presence of polyhydramnios. We decided the indications of the EXIT procedure by referring to the findings of prenatal imaging in the individual cases.

CHAOS is an extremely rare prenatally diagnosed clinical condition involving the complete intrinsic obstruction of the fetal airway [2]. Generally, the natural course of a fetus with CHAOS is fatal. The diagnosis of CHAOS based on prenatal imaging is established by bilateral enlarged echogenic lungs, dilated airways, flattened or inverted diaphragms, fetal ascites caused by fetal circulatory failure and hydrops [2]. Although the causes of CHAOS vary, the most common cause of CHAOS is laryngeal atresia [15]. In our series, the postnatal diagnosis of CHAOS was laryngeal atresia in all cases. Although all the cases with laryngeal atresia survived during the neonatal period, two cases died of respiratory failure in infancy. These cases had associated anomalies involving the respiratory tract and had a low birth weight. Thus, the prognosis of laryngeal atresia with isolated airway obstruction may be significant favorable, whereas the prognosis of patients with associated anomalies involving the respiratory tract may be poor. The EXIT procedure is a significant technique for safely performing tracheostomy in cases of laryngeal atresia and in improving the outcomes of such patients.

Although all the cases with CHAOS underwent the EXIT procedure, which led to the achievement of 6-month survival, during the follow-up period, the overall mortality rate of the CHAOS cases was 50%. In the last decade, isolated cases of CHAOS managed by fetoscopic decompression of the upper airway including laser laryngotomy have been reported [16, 17]. However, fetoscopic surgery is not approved for the treatment of CHAOS in Japan.

Fetuses with giant neck masses sometimes show marked the distortion or compression of the upper airways, and are in danger of life-threatening airway obstruction at birth. Adequate airway management improves the neonatal outcomes of such patients [5–9, 18]. These anatomical abnormalities of the fetal airways are diagnosed by prenatal imaging studies

before airway management. Fetal MRI can visualize the neck tumor and abnormal anatomy of the airways, and is especially helpful in the differential diagnosis of neck masses [19]. Airway patency is also evaluated by the fluid flow associated with fetal breathing on fetal ultrasonography. If the patency airway is confirmed prenatally, endotracheal intubation will be the first-line therapy for airway management. It is difficult to determine the indications for the EXIT procedure in cases involving fetuses with giant neck masses because it depends on the degree of upper airway compression by the mass. Hirose et al. proposed a management algorithm for cervical teratoma, recommending the EXIT procedure in fetuses with polyhydramnios and evidence of airway compression on fetal MRI [6]. In case 7, which involved a cervical teratoma that had expanded into the oral cavity, the patient died due to airway management failure as well as the unstable hemodynamics of the fetus during the EXIT procedure. We should have planned tracheotomy with the removal of the tumor from the anterior surface of the trachea as an alternative airway management in case of endotracheal intubation failure.

Even if the fetal airway could be successfully managed under the EXIT procedure, the patient's prognosis depends on tumor resectability. Only the cases in which the tumors were resected after birth survived to discharge. Although the patient in case 6, who had epignathus and was delivered at 28 weeks of gestation, could receive tracheostomy, a large amount of bloody effusion leaked directly from the tumor, resulting in death due to failure to achieve stable circulation. We should have planned some treatment options before birth, such as immediate resection of the tumor during the EXIT procedure or ligation of the tumor base after birth to avoid massive bleeding. Cervical teratoma usually has mixed components consist of a solid part and a cystic part [20]. In case 8, the patient with a cervical teratoma required immediate surgical resection because of the rapid enlargement of the cystic components of the tumor. We insist that before performing the EXIT procedure we should consider not only airway management at delivery, but also strategy for treating the tumor itself in cases involving a giant neck mass arising around the airway. One of the limitations was the retrospective study with a small number of patients. The other limitation of this study was the long study period, as there may be an institutional and clinical learning curve during this period. A large cohort and short study period would be necessary to investigate the differences in individual factors and evaluate the indications for the EXIT procedure in patients with fetal airway obstruction.

Conclusion

We concluded that the EXIT procedure was effective for the airway management of fetuses in which potential airway obstruction was prenatally diagnosed and that the procedure could be safely performed without any severe maternal complications. Especially in cases involving giant neck masses arising around the airway, airway management at delivery and the treatment strategy for the tumor itself should be planned before birth.

Compliance with ethical standards

Conflict of interest All authors declare that they have no conflict of interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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