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Exit to Airway: Delivery of a Term Fetus with Anterior Neck Mass

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Abstract – Ex-utero intrapartum treatment (EXIT) to airway is a rare and complex procedure performed during caesarean section, in which the fetal airway is secured before the umbilical cord is clamped to sustain uteroplacental circulation. It is a life-saving procedure used in cases of congenital upper airway anomalies or obstruction caused by cervical masses. It enables securing the fetal upper airway while still supported by placental circulation, thus decreasing hypoxia-related morbidity. It is relatively safe when executed by skilled and well-trained personnel. We performed EXIT to airway on a term baby with an anterior neck mass detected antenatally with ultrasonography. It was conducted smoothly with interdepartmental involvement including an obstetrician, a fetal-maternal team, neonatologists, and a paediatric otorhinolaryngologist, with well-structured plans and simulations. The ORL team performed direct laryngoscopy and assisted telescopic intubation, completing the procedure in approximately five minutes.

Keywords – EXIT to airway, Ex-utero intrapartum treatment (EXIT), uteroplacental support, cervical neck mass

1 INTRODUCTION

The Ex-utero intrapartum treatment (EXIT) was initially developed in the 1990s to remove tracheal occlusion devices placed during the intrauterine period for therapeutic purposes in cases of congenital diaphragmatic hernia (1,2). It was then gradually adapted to manage upper airway obstructions or cardiorespiratory anomalies where immediate intervention at birth was necessary to prevent morbidity and mortality (1,3-5). Different types of EXIT procedures are tailored to the indication and goal (1,4,6). EXIT to airway is the most documented procedure, followed by EXIT to resection. EXIT to resection includes excision of cervical, thoracic, or mediastinal masses that impinge on the airway (1,2,4). EXIT to extracorporeal membrane oxygenation (ECMO) is typically considered a last resort to support cardiopulmonary circulation, often in cases of congenital diaphragmatic hernia (CDH) (1-4). It remains a complex procedure due to its rarity and the associated morbidity for the mother (3,4). EXIT to airway is considered a game-changing procedure as it provides adequate time to establish a definitive upper airway by maintaining uteroplacental support for respiration and enables a smooth transition to extra-uterine life (3,7).

2 CASE REPORT

A 27-year-old G3P1+1 presented at 37 weeks, referred from primary care for an intrauterine growth restriction (IUGR) with an anterior neck mass for further investigation and management. She was admitted to the prenatal ward, and a detailed scan was performed. Ultrasound showed parameters at 29-33 weeks with a normal amniotic fluid index of 10 and an anterior neck mass measuring 4 x 3 cm with cystic consistency. Otherwise, there are no anomalies in the brain, spine, and heart, and the Doppler showed a normal waveform. Antenatally, she tested positive for IgG antibodies for toxoplasmosis and cytomegalovirus. She had one previous complete miscarriage 4 years before, which was not investigated. The second pregnancy was uneventful, and the child is healthy to date.

She was scheduled for a consultation with the maternal-fetal team for further evaluation and had received an intramuscular dexamethasone injection. However, she was in labour, and an emergency caesarean section was planned. The case was referred to the paediatric team to stand by for neonatal resuscitation and to otorhinolaryngology to secure the airway in view of an anterior neck mass.

Repeated ultrasound was performed, and the fetus was in a cephalic position with occiput posterior which was feasible for EXIT. Discussion and coordination done between anaesthetists, neonatologists, paediatric otorhinolaryngologists and obstetricians.

During the EXIT procedure, partial delivery of fetus where only the fetal head and neck were delivered through the hysterotomy. This limited exposure facilitated surgical access while minimizing heat loss. Direct laryngoscopy was then performed, and the fetal airway was secured by telescopic intubation with a 3.0 mm endotracheal tube (Figure 1 and 2). Direct laryngoscopy findings show the base of the tongue normal, not prolapsing posteriorly; epiglottis and arytenoid mucosa normal, not prolapsing into the airway; bilateral vocal cords symmetrical; no visible mass or medialisation of the airway, with no evidence of tracheal stenosis or laryngeal cleft. Intubation was smooth with a single attempt, anchoring at 9 cm. The baby was born weighing 1635g with an Apgar score of 5T, a heart rate of 150 beats per minute with SpO₂ ranging 85-92%, and neonatal resuscitation was undertaken. The EXIT procedure lasted about five minutes, while the caesarean section took approximately 45 min.

On examination, the mass is located more over the right lateral neck, extending from the angle of the mandible down to the inferior third of the sternocleidomastoid muscle, anteriorly up to the thyroid cartilage, and posteriorly extending to the anterior border of the trapezius. The mass is cystic in nature and measures approximately 5 x 3 cm (Figure 3). Unfortunately, syndromic features observed upon examination of the baby include low-set ears, widely spaced nipples, a fixed deformity over the left hand with clinodactyly, and preaxial deformity of the left thumb. Bedside echocardiography suggests a complex cyanotic heart disease. Fluorescence in situ hybridisation (FISH) was done to confirm Edward syndrome. The baby was extubated on day 4 of life and able to wean off oxygen, discharged home with palliative intent.

3 DISCUSSION

The EXIT procedure is a complex and specialised intervention that is rarely performed but is designed and adapted to provide sufficient time to secure the fetal airway while maintaining uteroplacental circulation for gas exchange. EXIT aims to secure the airway through conventional or assisted intubation or tracheostomy when intubation is not possible (1-3,6).

Indications for EXIT to airway include congenital airway anomalies or a large cervical mass which can impede the upper airway (3,4,7). It is generally categorised into extrinsic and intrinsic factors (1,3). The most common causes of extrinsic compression include teratomas in the neck or pharyngeal region, which pose the highest risk due to their location (3,4). Vascular malformations, such as haemangiomas, which are the most common neonatal soft tissue tumours, followed by cystic lymphatic malformations, congenital giant ranula, fetal goitre, micrognathia, and others, can also cause extrinsic compression (2-4). In our case, it is an extrinsic compression by an anterior cystic neck mass, likely a cystic hygroma, measuring 4 x 3 cm (Figure 3).

Obstructions at lower levels of the airway, caused by large thoracic masses, may require alternative EXIT procedures designed to relieve upper airway obstruction. The severity of airway impairment mainly depends on the size, consistency, and location of the mass. Intrinsic factors include congenital high airway obstruction syndrome (CHAOS), a rare anomaly characterised by the absence of canalisation of the upper airway during the first trimester (1,3,4). This syndrome may involve conditions such as subglottic stenosis or atresia, laryngeal agenesis, or webbing.

Due to the rarity of these procedures, performing them in an emergency setting will increase maternal morbidity and risk due to a lack of preparation and untrained personnel (2,3,7). Ensuring stable maternal circulation is crucial to maintaining uteroplacental circulation, which allows more time to secure the airway for ventilation, thereby facilitating the fetus's adaptation to extrauterine life and preventing hypoxic-related morbidity (1,3). In this case, no fetal lung or diaphragm anomalies are identified, and the mass is not larger than 5 cm. Only assisted intubation is anticipated, and a short duration of EXIT to airway is required. Despite referral during active labour in emergencies, management at the tertiary centre is supported by a skilled multidisciplinary team, including paediatric otorhinolaryngologists, obstetricians, anaesthetists, paediatricians, and neonatologists (4). This allows for discussion, structured planning, and the elective execution of the EXIT procedure with a tailored algorithm (4,6). Success rate of EXIT to airway is directly related to early referral to a tertiary centre with maternal-fetal specialists, as it allows sufficient time for investigation and planning (2,4).

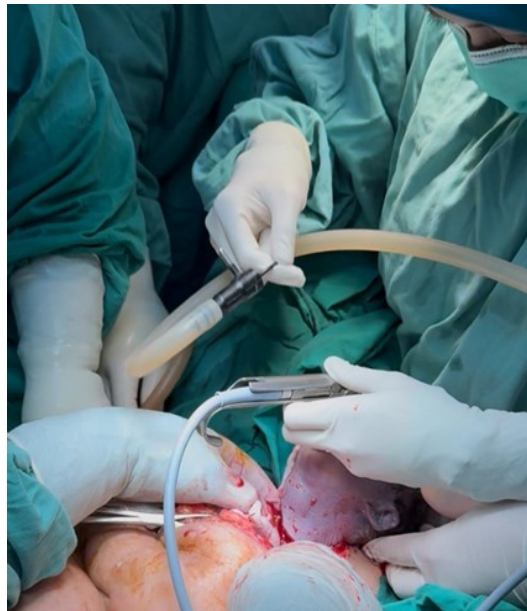


Figure 1. Direct laryngoscopy by using Parson's laryngoscope during EXIT procedure. Only the head and neck were extracted to allow the EXIT procedure

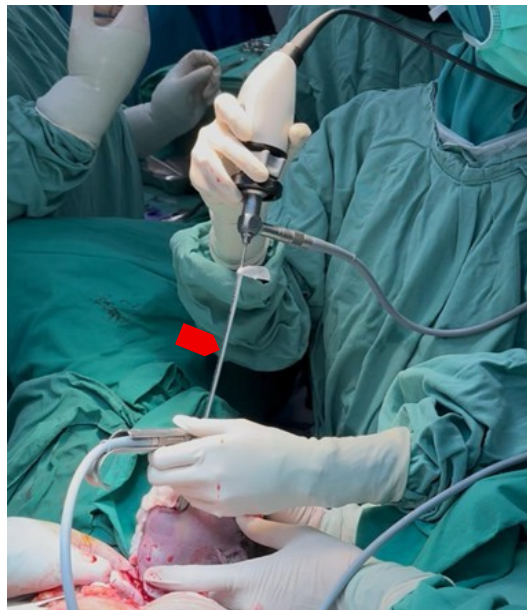


Figure 2. Telescopic intubation with endotracheal tube (ETT) sleeved over rigid endoscope rod (red arrow)



Figure 3. Anterior neck mass measuring 5 x 3 cm (red arrow)

Prenatal assessment by obstetricians and the maternal-fetal team, including detailed scans, can provide information on placental location, amniotic fluid index, airway anomalies, diaphragm position, and cardiac anomalies (1,3,4). The detection of a cervical mass during a detailed scan should include its size, whether it is cystic or firm, its vascularity, and its location (midline anterior or lateralised) (1,2,6). Magnetic resonance imaging (MRI) provides more information about the mass's characteristics than ultrasonography during a detailed scan by an obstetrician. It can also show its relationship to nearby structures, including any displacement of the aerodigestive tract (5,7). However, MRI was not performed in our case as the patient was already in labour. Furthermore, ultrasound assessment by an obstetrician is sufficient when there is evidence of a patent airway. Radiological imaging is essential in planning the EXIT to airway procedure by providing information on the size and consistency of the mass, airway patency, and structural abnormalities, which assists otorhinolaryngologists choose the best approach to secure the airway (1,3,6).

In a large mass compressing the airway with minimal displacement, direct laryngoscopy endotracheal intubation can be attempted. If it fails, the procedure proceeds with rigid bronchoscopy with telescopic-assisted intubation. If CHAOS is diagnosed prenatally, tracheostomy should be performed (1,3,4,6). Ideally, if prenatal imaging suggests a syndromic fetus, genetic studies should be performed to stratify fetal prognosis, and EXIT procedures should be avoided if congenital anomalies indicate a high risk

of non-viability after birth, as the procedure increases maternal risks with no neonatal benefit (2). However, prenatal genetic testing is not readily available in most healthcare facilities in Malaysia, where multidisciplinary teams rely mainly on fetal ultrasound to guide decisions in addressing airway obstruction in emergency situations. The postnatal diagnosis of Edwards syndrome in our case highlights the diagnostic gap that can arise. This also highlights the importance of improving access to genetic screening as a prenatal assessment, which could be helpful in parental counselling and support informed decisions on complex procedures such as EXIT.

Up to current practice, there are no guidelines on selecting cases for the EXIT to airway procedure, but it should be considered in any cases of potential airway obstruction (4,7). Commonly used guides include considering a cervical mass size greater than 3.5cm, some suggest 5cm as a cutoff point for an EXIT to airway procedure (2,3,6,7). Smaller sizes should also be considered if there is evidence of airway compression suggested by imaging (7).

A multidisciplinary team discussion is essential to ensure the smooth conduct of the procedure and to minimise morbidity for both mother and fetus. The team should comprise an obstetrician, maternal-fetal specialists, anaesthetists, neonatologists, and paediatric otorhinolaryngologists (2-6). Elective caesarean section is ideally planned close to 37 weeks to ensure fetal lung maturity and avoid preterm labour, thus reducing maternal and fetal morbidity (1,3). The obstetrician performs detailed scans to assess fetal position, placental location, and neck

mass characteristics, followed by counselling on procedural risks and implications for future pregnancies (1,4,6,8). Two anaesthetic teams were involved: the maternal team was responsible for uterine relaxation to maintain uteroplacental perfusion, while the fetal team was responsible for fetal monitoring (2-4,6). Paediatric otorhinolaryngologists assess airway patency, mass size and characteristics to determine the best way to secure the airway by direct laryngoscopy intubation, assisted intubation, or a tracheostomy required (3,4,6). The fetal position determines the degree of exposure for airway manipulation and influences the complexity of the EXIT procedure, where cephalic and occiput posterior are ideal (3,6). At least one multidisciplinary simulation is recommended to ensure coordinated execution of the EXIT procedure where all team members familiarize with their role and algorithm (3,4). All instruments and equipment are prepared in advance according to fetal weight and gestational age (3,4,6).

The proposed setup in the operating theatre where we incorporate in our case is the pregnant mother lies with her head towards the anaesthetic circuit in the lithotomy position. The mother's anaesthetist team will be at the head of the mother, while obstetricians will be on her right side to perform the caesarean section, with a scrub nurse on her left. Paediatric otorhinolaryngologists, as the lead person to secure the fetal airway, should be positioned between the mother's legs for easy airway manipulation, and the fetal anaesthetist team will be over the lower right side of the mother (1,3). The neonatologist should be close to the paediatric otorhinolaryngologist to take over resuscitation once the airway is secured (4). An overhead warmer is placed at the end of the operating table.

During the EXIT procedure, only the fetal head up to the upper chest should be exposed to provide the necessary surgical field and ensure full visualisation of the cervical mass, while allowing proper positioning of the neck for intubation (1,3,4). We should limit fetal exposure to prevent heat loss and hypothermia (3,4,6). In our case, the obstetricians performed a standard hysterotomy without extending the incision, extracting only the head and neck, as this provided sufficient access to visualise the mass without needing to expose the upper chest. One arm was also delivered for vital signs monitoring by the anaesthetists. The duration of the exit procedure usually ranges from 5 to 22 minutes (2,3).

In our case, less than 5 minutes are required to secure the fetal airway. Direct laryngoscopy is performed, and telescopic intubation proceeds without difficulty. We chose telescopic intubation as we can perform an airway study at the same settings. Neonatologists will observe fetal perfusion, muscle tone, and oxygenation.

4 CONCLUSION

Given the complexity of the delivery process and rarity of these procedures, there are no established guidelines on indications, algorithms, and contraindications for the EXIT procedure. This case highlighted the importance of early referral to a tertiary centre as it is equipped with skilled personnel from all the disciplines involved. Clear communication will ensure success of the EXIT procedure. A tailored algorithm should be developed for each case to ensure airway is secured in the shortest possible time during elective procedures.

AUTHOR CONTRIBUTIONS

Low SW: Drafting of the article, Collection and assembly of data, Final approval of the article. Aziz A: Analysis and interpretation of the data, intellectual content, Final approval of the article. Mohamad I: Conception and design, Critical revision of the article for important revisions, Final approval of the article.

AUTHORSHIP CLARIFICATION

All authors meet the ICMJE criteria for authorship and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

No ethical approval was necessary for this case report under our institutional policy. Informed consent was obtained from the participant.

CONSENT FOR PUBLICATION

Informed consent was obtained from the patient for publication of the case and the images.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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