

A Five-year Experience of EXIT Procedure in East Coast of Malaysia: A Literature Review with Cases

Malezya'nın Doğu Kıyısında Beş Yıllık EXIT Prosedürü Deneyimi: Vakalarla Bir Literatür İncelemesi

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ABSTRACT

Introduction: With the advancement of the prenatal assessment and early diagnosis of foetal head and neck lesions in utero that potentially causing upper airway obstruction in neonates, Ex Utero Intrapartum Treatment (EXIT) procedure has becoming an accepted, updated medical intervention worldwide to manage such cases. This method has led to a significant change in neonatal management and improved neonatal outcomes. The objective of this study was to describe and evaluate the 5-year experience with EXIT procedure in the year 2014 till 2019 at the East Coast region in Malaysia from the paediatric ENT perspective.

Method: All EXIT procedures performed in 2 hospitals in East Coast region of Peninsular Malaysia, which are Hospital Raja Perempuan Zainab II and Hospital Sultanah Nur Zahirah between 2014 and 2019 were retrospectively reviewed. We study EXIT Procedures performed during that period, focusing on the prenatal assessment, pre-operative planning, intraoperative and post-operative management of the cases. We also analysed the postoperative outcome of the patients.

Results: EXIT procedures were performed in five cases with prenatal diagnosis of cystic hygroma in three cases, one case of epulis and one case of Congenital High Airway Obstruction Syndrome (CHAOS). Airway management was successful with endotracheal intubation in 4 cases (three cases of cystic hygroma and one case of epulis) and failed in the CHAOS case. The CHAOS case underwent tracheostomy after bronchoscopy but succumbed to the disease. Long term survival was achieved with the four successful EXIT cases.

Conclusion: Results from our series of EXIT procedure was effective and can be performed safely in the airway management of fetuses suspected airway obstruction. Early referral and multidisciplinary teams involvement and planning were very crucial for a planned delivery and should be done earlier before birth.

Keywords: EXIT procedure, ex utero intrapartum treatment, CHAOS, cystic hygroma, congenital high airway obstruction syndrome

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ÖZET

Giriş: Yenidoğanlarda potansiyel olarak üst hava yolu obstrüksiyonuna neden olan utero fetal baş ve boyun lezyonlarının prenatal değerlendirme ve erken teşhisinin ilerlemesiyle birlikte, Ex Utero Intrapartum Treatment (EXIT) prosedürü, bu tür vakaları yönetmek için dünya çapında kabul görmüş, güncellenmiş bir tıbbi müdahale haline geldi. Bu yöntem, yenidoğan yönetiminde önemli bir değişikliğe ve yenidoğan sonuçlarının iyileştirilmesine yol açmıştır. Bu çalışmanın amacı, Malezya'nın Doğu Kıyısı bölgesinde 2014'ten 2019'a kadar olan 5 yıllık EXIT prosedürü deneyimini pediatrik KBB perspektifinden tanımlamak ve değerlendirmektir.

Yöntem: 2014-2019 yılları arasında Malezya Yarımadası'nın Doğu Kıyısı bölgesindeki 2 hastanede Raja Perempuan Zainab II Hastanesi ve Sultanah Nur Zahirah Hastanesinde gerçekleştirilen tüm EXIT işlemleri geriye dönük olarak incelendi. Bu dönemde gerçekleştirilen EXIT Prosedürlerini, doğum öncesi değerlendirme, ameliyat öncesi planlama, vakaların ameliyat sırasında ve ameliyat sonrası yönetimine odaklanarak inceliyoruz. Ayrıca hastaların ameliyat sonrası sonuçlarını da inceledik.

Bulgular: Prenatal kistik higroma tanılı beş olguya, üç olguya, bir olguya epulis ve bir olgu Konjenital Yüksek Havaolu Tıkanıklığı Sendromu (CHAOS) tanısı ile EXIT işlemi uygulandı. Hava yolu yönetimi, 4 vakada (üç kistik higroma ve bir epulis vakası) endotrakeal entübasyon ile başarılı oldu ve CHAOS vakasında başarısız oldu. CHAOS olgusuna bronkoskopi sonrası trakeostomi açıldı ancak hastalığa yenik düştü. Dört başarılı EXIT vakası ile uzun süreli sağkalım sağlandı.

Sonuç: Hava yolu obstrüksiyonundan şüphelenilen fetüslerin hava yolu yönetiminde EXIT prosedür serimizin sonuçları etkiliydi ve güvenle uygulanabilir. Erken sevk ve multidisipliner ekiplerin katılımı ve planlaması, planlı bir doğum için çok önemliydi ve doğumdan önce yapılmalıdır.

Anahtar Sözcükler: EXIT prosedürü, ex utero intrapartum tedavi, CHAOS, kistik higroma, konjenital yüksek hava yolu obstrüksiyonu sendromu

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INTRODUCTION

Ex Utero Intrapartum Treatment (EXIT) procedure is a technique to manage foetal airway in cases suspected to have airway obstruction while maintaining uteroplacental circulation to the foetus with neonatal anaesthesia by controlled uterine hypotonia (1-8). The indications include congenital neck masses such as cystic hygroma and congenital high airway obstruction syndrome (CHAOS).

The advancement in prenatal diagnostic and foetal imaging including ultrasonography and magnetic resonance imaging (MRI) has led to early detection and diagnosis of potential neonatal airway obstruction at birth. This allows and opens an opportunity for airway management and prevention prior to the delivery.

METHODS

This is a retrospective case review of five consecutive experience of EXIT procedures that has been performed in East Coast region of Peninsular Malaysia. This study involved two hospitals, which are Hospital Raja Perempuan Zainab II and Hospital Sultanah Nur Zahirah between 2014 and 2019. The airway management involved foetal airway obstruction with three cases of cystic hygroma, one case of epulis and one CHAOS. The maternal and infants' medical records were retrospectively reviewed to obtain the demographic data. The maternal data obtained includes maternal age, gravidity and parity, gestational age at initial diagnosis, the prenatal diagnosis and findings of foetal imaging or foetal MRI. The neonatal data included gestational age at delivery, sex, birth weight, associated anomalies and outcome were analysed.

Ex utero intrapartum treatment

For all five cases, the initial prenatal diagnosis was made during prenatal ultrasonography in scheduled antenatal follow up in the community clinics. These cases were subsequently referred to tertiary centre for further investigations including MRI, follow up and planning. Multidisciplinary team joint involvement and conference held to discuss and planning for the delivery, the anaesthesia method, and the optimal methods of airway management. The multidisciplinary teams for EXIT procedures performed consist of obstetricians, anaesthesiologists including paediatrics anaesthesiologists, paediatrics otolaryngologists, neonatologists and operating room nurses. The detail planning of EXIT procedure has to be individualised based on the case.

In all of our cases, EXIT procedures were conducted as elective cases and at full term, as the mother and foetus were in stable conditions. However, detail planning and preparation were also outlined as preparation if the conditions became unstable and mothers need to be pushed to theater for emergency procedure. The operation theater arrangements and organizations were done as planned.

The EXIT procedures were performed during caesarean section under general anaesthesia. Once the foetal head and neck (up to one of shoulder) exposed, the airway can be secured either via endotracheal intubation or tracheostomy. During EXIT procedure, all babies were in supine position and the rest of the foetal body remained in utero.

Once the airway secured, manual ventilation was initiated by the paediatrics anaesthesiologist while still on uteroplacental circulation. The neonate then fully delivered, umbilical cord clamped and cut. The neonate transferred to neighbouring operation theatre for further assessment and resuscitation. The maternal uterus and abdomen closed.

RESULTS

The demographic data of the mothers

Total of five cases of EXIT procedures has been performed in five years (Table 1). The maternal age ranging from 20 to 36 years old, with median age of 27.2 years. The median and range of gestational age at initial diagnosis was 28.2 (20-32) weeks of gestation. All of the cases initially diagnosed during foetal ultrasonography and subsequently further assessed with foetal MRI. Three of the foetus diagnosed with cystic hygroma, one case of epulis and one case of CHAOS.

Table 1: The demographic data of the mothers and prenatal diagnosis

Case No	Maternal age	Gravidity and parity	Gestational age at initial diagnosis (week)	Prenatal diagnosis	Findings of fetal imaging (MRI)
1	36	G4 P3	32	Cystic hygroma	6 x 3 cm Cystic hygroma, fixed flexion of fingers, echogenic bowel
2	25	G2 P1	34	Cystic hygroma	6.6 x 6.3 x 3.7 cm multiloculated anterior neck swelling, solid mixed with cystic component, with no obvious narrowing/ external compression
3	33	G5 P1+3	20	CHAOS, left lung dysplasia and tetralogy of fallot	Fetal CHAOS with typical hyperechoic lungs, everted diaphragm, dilated trachea and ascites with atretic site locates at subglottic area. Also has tetralogy of fallot with right aortic arch
4	20	G1 P0	35	Fetal oral tumour	28 x 24 x 20mm oral tumour, homogenous solid in nature arising from lower anterior and behind the lower lip
5	22	G1 P0	20	Cystic hygroma	10.9 x 24.8 x 19.5cm multilobulated, multiseptated cystic neck lesion with acute-early subacute bleed

Summary of the EXIT procedures

The gestational age at delivery ranging from 37-38 weeks with median of 37.6 weeks (Table 2). In all cases, foetus was stable hemodynamically during EXIT procedure.

As for our first and second EXIT procedures, intubation using glide scope failed on the first attempt due to oedematous epiglottis (case 1) and unable to identify epiglottis (case 2). In both cases, the laryngeal mask airway (LMA) size 1 was inserted, then the endotracheal tube size 3 was inserted via rail route technique through flexible fiberoptic bronchoscope by the paediatrics otolaryngologist.



Picture 1: Endoscopic view of the laryngeal structures post intubation (case 2)

For our third case, rigid bronchoscopy done confirmed tracheal stenosis of grade 4 (Cotton Myer classification)(9). Tracheostomy then performed and tracheostomy tube size 3 inserted without difficulty. However, highest oxygen saturation achieved was 40%. Unfortunately, with full resuscitation, the neonate succumb to bradycardia and desaturation.

As for the fourth and fifth case airway management, both foetus managed to be intubated at first attempt via direct laryngoscopy. The procedures were uneventful.



Picture 2: Case 4 after intubation during EXIT procedure



Picture 3: Picture of Case 5 after intubation

Table 2: Summary of the EXIT procedures

Case No	Timing of EXIT	Gestational age at delivery (week)	Stability of foetal hemodynamics	Airway managements	Postoperative complications
1	Elective	37	Stable	LMA, Fibreoptic bronchoscopy guided endotracheal intubation	None
2	Elective	38	Stable	LMA, Fibreoptic bronchoscopy guided endotracheal intubation	None
3	Elective	37	Stable	Tracheostomy	Death
4	Elective	38	Stable	Direct laryngoscopy endotracheal intubation	None
5	Elective	38	Stable	Direct laryngoscopy endotracheal intubation	None

Outcome of the foetuses

The postnatal diagnosis in three of the cases were cystic hygroma, one case of epulis with histological result of granular cell myoblastoma and one case of tracheal stenosis of grade 4 (total occlusion of lumen).

We had long term survival of 4 cases. For case 1, tracheostomy was done at day 22 of life and then subsequently debulking of the cystic hygroma done at day 55 of life. In case 2, tracheostomy and debulking was done earlier, which was at day 8 of life. Case 4 did not require tracheostomy and resection of tumour was performed on day 4 of life. As for our fifth case, the debulking of cystic hygroma on day 8 of life and did not required tracheostomy.

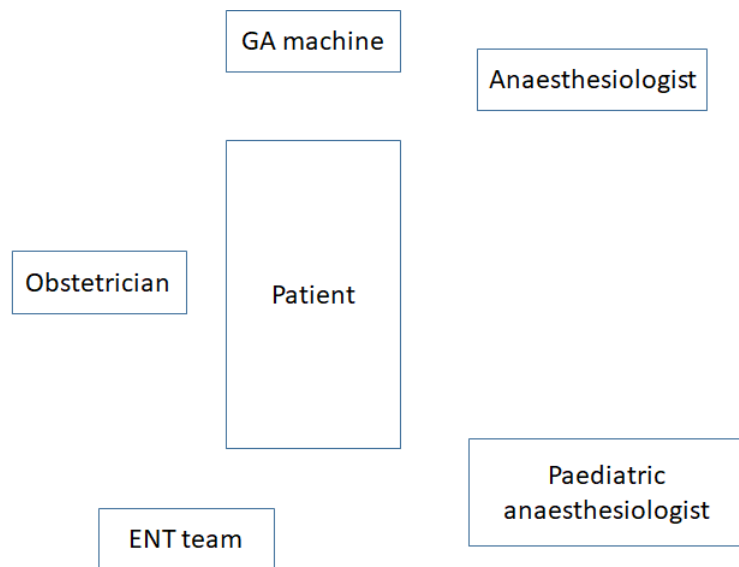
Table 3: Outcomes of the foetuses

Case No	Postnatal diagnosis	Sex	Birth weight (g)	Associated anomalies	Outcome
1	Cystic hygroma	Boy	3500	None	Alive
2	Cystic hygroma	Girl	2800	None	Alive
3	Tracheal stenosis (Grade 4)	Boy	2950	Left lung dysplasia, Tetralogy of fallot	Death
4	Epulis	Girl	2600	None	Alive
5	Cystic hygroma	Boy	5000 (estimates fetal weight 2.2kg)	None	Alive

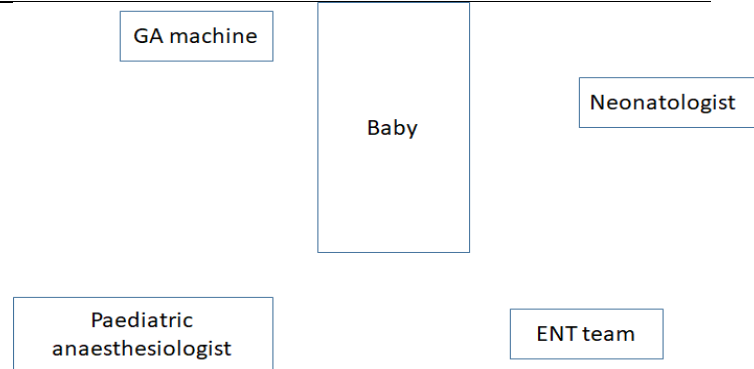
DISCUSSION

Early, prenatal diagnosis of head and neck masses during antenatal allow paediatrics otolaryngologists to secure the foetus airway while maintaining the uteroplacental circulation during caesarean section(10). This advancement much supported by the presence of prenatal ultrasonography and availability of foetal MRI. The whole procedure commonly done under general anaesthesia to achieve uterine relaxation⁸ and this is the basis of EXIT procedure performed. During EXIT procedure, head and one of the foetal shoulder are delivered while maintaining the remaining part of the foetal body and the umbilical cord inside the uterus. These help to preserve heat as well as blood loss(11,12).

Here, we shared the operation theatre arrangement for the EXIT procedure.



Graphic 1: Set up of the operation theatre during EXIT procedure.



Graphic 2: Arrangement of the paediatric operation theatre after EXIT procedure.

There are few practical tips that we learnt during the procedures that help in smooth out and ease the process. OT running EXIT procedure is always cramped with multiple team members from various departments. Each team members must be labelled accordingly (Picture 4) to ensure smooth flow of the procedure. Labelled trolley(Picture 5) and instruments are prudent to make sure members do not fight for same instrument (eg: suction machine)causing delay especially during critical period .



Picture 4: Colour coded label for the team members



Picture 5: Well-arranged ENT instruments

In our experiences, four out of five cases had successful intubations. We would like to highlight the intubation process of case 1 and case 2 as an alternative in a failed direct laryngoscopy intubation apart from intubation using rigid bronchoscope. For both cases, on initial attempt using direct laryngoscopy failed as we had difficulty to identify the laryngeal inlet. LMA size 1 was then inserted to first secure the airway. After that, the endotracheal tube size 3 was inserted via rail route technique through flexible fiberoptic bronchoscope. After successful intubation of the neonate, LMA was removed. Similar intubation technique was applied in case 1 and 2.

In case 3, with a rare diagnosis of CHAOS, which was diagnosed prenatally and detail planning of EXIT procedure, we lost the neonate to respiratory failure. Despite successful tracheostomy, the anomalies of the respiratory tract and lung gave rise to poor prognosis. As for case 4 and 5, endotracheal intubation successfully done via direct laryngoscope.

All of the four successful cases, the management followed by resection of the tumour. Two out of the four cases require tracheostomy due to extensive and anteriorly located lesion.

CONCLUSION

This study is first to evaluate the EXIT procedures and the foetuses outcome for the cases performed in East coast of peninsular Malaysia. The EXIT procedure was effective to manage airway for suspected airway obstruction prenatally. It is a safe procedure but require the involvement of multidisciplinary team, joint conferences and detail planning before the delivery of foetus.

Conflict of interest

No conflict of interest was declared by the authors.

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