Ex-utero intrapartum treatment: a controlled approach to the management of anticipated airway problems in the newborn

Airway problems in an unborn foetus that may cause obstruction can be safely managed using an ex-utero intrapartum technique. Advanced technology now allows many congenital airway problems to be diagnosed in the prenatal period. Careful prenatal planning of an ex-utero intrapartum treatment allows safe airway control while the foetus remains on uteroplacental support. It avoids the need for emergent intervention of an acutely obstructed airway in a neonate that often has disastrous consequences.

Introduction

The ex-utero intrapartum treatment (EXIT) procedure was initially developed for the removal of a tracheal clip placed to create tracheal obstruction in a foetus with severe congenital diaphragmatic hernia. It is now established as a safe technique for the management of potential foetal airway obstruction. The principle behind the use of tracheal occlusion in congenital diaphragmatic hernia is to obstruct lung fluid circulation, stimulating lung expansion. To maintain uteroplacental circulation during the intrapartum period, the uterus must be kept in a relaxed state. As a result, the foetus is maintained on placental circulation while the tracheal clip is removed. The EXIT procedures have been proven effective for the management of foetal airway obstruction due to a variety of causes. These include neck masses that may cause airway obstruction, congenital high airway obstruction syndrome (CHAOS), intrathoracic masses, and congenital diaphragmatic hernias. The EXIT procedure allows sufficient time for the establishment of an airway via intubation, tracheostomy, and even resection of the obstructing lesion with the baby on placental support. The reported duration for EXIT is usually around 1 hour, although it may be up to 2.5 hours.

Case report

A 36-year-old woman in her second pregnancy was referred to our prenatal clinic for prenatal diagnosis because of advanced maternal age. She elected to undergo amniocentesis. The ultrasound examination of foetal morphology during amniocentesis at 18 weeks’ gestation showed a 3-cm soft tissue mass situated at the anterior chest wall, right arm, and neck. Amniocentesis was uneventful and the provisional diagnosis of a subcutaneous haemangioma / lymphangioma was made. Serial ultrasound examinations performed at 2- to 3-week intervals revealed a progressive increase in the lesion size: by 32 weeks’ gestation the greatest diameter of the mass was 10 cm (Fig 1). The foetal growth variables were otherwise normal. Chromosomal analysis revealed a normal 46XY karyotype.
In view of the large anterior chest wall mass, the possibility of airway obstruction was raised. Detailed ultrasound assessment at that time, including attempts to use real-time three-dimensional ultrasound, showed no evidence of polyhydramnios and no disfigurement of the foetal face. Nonetheless distorted neck posture due to the presence of the large mass was evident. Foetal magnetic resonance imaging was not performed as the relevant expertise and experience in this area were unavailable. The ENT surgeon, neonatologist, anaesthesiologist, and paediatric surgeon discussed possible airway intervention during an elective delivery by caesarean section. The consensus was that there was sufficient concern about airway obstruction at the time of delivery to warrant additional precautions. The EXIT procedure with different possibilities of airway intervention was discussed in detail with the parents. The parents accepted the recommendations of the multidisciplinary team, despite the understanding that this would be the first fully monitored EXIT in Hong Kong. Caesarean section with EXIT procedure was scheduled for 36 weeks of gestation. It was agreed that should spontaneous labour occur before this time and emergency Caesarean delivery be needed, the EXIT procedure might not be feasible and the presence/absence of airway obstruction could only be assessed during neonatal resuscitation.

The operation was performed in May 2005 as the first case of the day by a team of experienced obstetric surgeons, ENT surgeons, anaesthesiologists, neonatologists, with two separate groups of scrub nurses. The theatre was divided into the maternal team and the foetal team.

The mother was put in a supine position on the operating room table, using a slight left lateral decubitus position to reduce aortocaval compression. Preoperatively, indomethacin had been prescribed as a tocolytic and general anaesthesia was administered in the standard fashion. High-dose inhalation agents and intravenous nitroglycerin were used to maintain uterine relaxation. The foetal heart rate and maternal oxygen saturation and blood pressure were monitored and well maintained intrapartally to ensure adequate uterine perfusion. After anaesthesia was induced, a laparotomy was performed in the standard fashion with Kerr’s incision made in the lower uterine segment. The position of the placenta was identified and mapped. The hysterotomy was planned to ensure that incision of the placenta could be confidently avoided. The uterus was opened and the foetal head was exposed (Fig 2a). The edges of the uterine wound were not plicated as bleeding was deemed not excessive.

The head and the left arm of the baby were delivered out of the uterus and a neonatal pulse oximetry sensor attached to the baby’s left hand. The monitor was covered with sterilised tin foil to avoid light interference from theatre lamps. Foetal pulse oximetry was maintained at around 50 to 60%. The 5th percentile for foetal pulse oximetry is reported to be 30% in normal uncomplicated labours and deliveries and persistent oximetry of less than 30% for 10 minutes in the foetus is associated with foetal acidemia. The foetal heart rate and placental circulation were monitored via continuous ultrasound Doppler examination. The ultrasound probe was draped with a sterile cover and placed on the maternal abdominal wall above the laparotomy incision (Fig 2b).

The upper airway was carefully examined with the help of a Benjamin slotted laryngoscope and a 4-mm rigid endoscope. The vocal cords were in the paramedian position. No obvious obstruction was visualised at the supraglottic level with the baby’s head turned to the neutral position. Nonetheless intubation with a size 2.5 Portex endotracheal tube was performed to ensure a secure airway (Fig 2c). The rest of the foetal trunk was delivered with the umbilical cord clamped and cut. The placental by-pass time was 24 minutes.

The baby was handed to the attending neonatologist for further assessment. Oxytocin was administered once the umbilical cord was clamped to prevent uterine atony and postoperative bleeding. The total maternal blood loss during the caesarean section was 400 mL. The whole operation time took 1 hour 17 minutes.

The airway of the baby was checked endoscopically and he was extubated successfully 2 hours following the EXIT procedure. The other systems were unremarkable. The mother had an uneventful postnatal recovery and was discharged on day 6.

Discussion

The EXIT procedure was originally designed to remove tracheal clips, positioned in utero to occlude the trachea in
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foetuses suffering from congenital diaphragmatic hernia. Placental circulation is maintained in this situation in order to allow time for clip removal, intubation or tracheostomy.

In addition to the above, the EXIT procedure is a valuable means of managing anticipated airway obstruction. Some common indications include a neck mass that obstructs the upper airway,^6^ CHAOS^7^, and thoracic abnormalities (eg hydrothorax).^8^

More rare types of congenital neck mass include cervical teratoma and cystic hygroma. Endotracheal intubation at birth can be very difficult in such cases: overall infant mortality is estimated to be 20% if the foetus is delivered vaginally. If the foetus is delivered via the EXIT procedure, intubation, tracheostomy or even resection of the lesion can be performed while the infant remains on placental support. The maximal duration of the EXIT procedure performed for excision of teratoma has been reported to be 2.5 hours.^3^

Congenital high airway obstruction syndrome is a spectrum of anomalies that includes the presence of laryngeal web, atresia, or cyst and tracheal atresia or stenosis. It is a rare abnormality of unknown cause and is characterised by enlarged lungs, dilated distal airways, everted diaphragm, ascites and, ultimately, non-immune hydrops foetalis. In such cases, it is much safer to establish airway control with EXIT and later correct the defect via a planned elective procedure once neonatal status is optimised.

Common types of intrathoracic abnormalities include congenital pleural effusion and pulmonary agenesis. Prontera et al^8^ described a foetus with bilateral pleural effusions that were drained at birth during an EXIT procedure. It allowed sufficient time for drainage of the effusions and lung expansion. A foetus with severe congenital cystic adenomatoid malformation can also be treated by EXIT for airway support and even resection of the cystic lung segment.^10^

A recent review of the EXIT procedure^2^ examined 52 cases performed between 1993 and 2003. Indications were congenital diaphragmatic hernia 87%, neck mass 10%, and CHAOS 3%. The gestational age at birth was 32 ± 3 weeks and time on placental support was 45 ± 25 minutes.

One of the major theoretical risks of the EXIT procedure is excessive maternal bleeding from the uterine incision. The use of surgical staplers to secure haemostasis

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Fig 2. Clinical photos showing (a) exposure of foetal head during the ex-utero intrapartum treatment, (b) ultrasound Doppler scan for monitoring foetal heart rate and placental circulation, (c) direct laryngoscopy and intubation, and (d) the foetus immediately after birth.

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(a) (b)
(c) (d)

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after delivery of the foetal head has been advocated. If appropriate staplers are unavailable, plication of the uterine incision can also be performed, but is more tedious and can interfere with airway management of the foetus. Major maternal haemorrhage has not been reported to date. In a recent series, the short-term maternal outcome following the EXIT procedure, including postoperative hospital stay and maternal haematocrit, was similar to that of mothers following routine caesarean section. The only difference was the higher rate of wound complications and estimated blood loss in the EXIT group.11

Foetal intervention does not adversely affect long-term maternal fertility and no maternal death has been reported.12

Conclusion

The success of EXIT depends on close cooperation in a multidisciplinary team that includes obstetricians, neonatologists, anaesthesiologists, ENT surgeons, and paediatric surgeons. It is a particular challenge for the anaesthesiologist who is responsible for maintaining uteroplacental circulation, whilst keeping the uterus relaxed. The haemodynamic status of the mother and foetus must be carefully monitored and titrated.

The EXIT procedure can be safely performed in foetuses with congenital diaphragmatic hernias, neck masses, and thoracic masses. With a meticulously planned protocol set up within the hospital, it obviates the need for emergency endotracheal intubation or tracheostomy in infants with high-risk airways, a situation that can have disastrous consequences.

References