Difficult neonatal airway in the context of an EXIT Procedure – Case Study

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Introduction

This case study describes the recent involvement of a Royal Children’s Hospital (RCH) anaesthesia team in a planned EXIT procedure performed at the Mercy Hospital for Women (MWH) in Melbourne. RCH is a quaternary paediatric hospital, and the MWH is a tertiary obstetric hospital both located in Melbourne.

EXIT stands for EX utero Intrapartum Treatment. It is a paediatric procedure, most commonly for airway compromise, performed on a neonate as the head and shoulders are delivered during Caesarean Section, while the neonate is still oxygenated via placental blood flow (Hirose & Harrison, 2003).

Expert paediatric anaesthetic assistance is required for this procedure due to the antepartum in utero diagnosis of congenital neonatal airway obstruction – likely to cause complete airway obstruction and death upon delivery with separation from the placental blood supply (Suenaga et al., 2014).

Background

The EXIT procedure is a rare surgical procedure performed to establish neonatal airway patency in situations of severe or life threatening airway obstruction. EXIT was developed in 1989 as part of the initial treatment of congenital diaphragmatic hernias, but is now been adapted to other congenital neonatal airway such as large neck masses, high airway obstruction and the separation of conjoined twins (Mayer et al, 2011).

The EXIT procedure allows the congenital airway issue to be addressed in a controlled way, while preserving the placental perfusion of the partially delivered neonate (Hirose & Harrison, 2003).
Case Study

In this case the EXIT procedure was indicated by the antenatal diagnosis of a lymphatic malformation in the head and neck of the neonate, causing a large neck mass with probable airway compromise. The mother had previously had a vaginal delivery of a healthy baby. For the pregnancy she was booked to deliver by elective caesarean section at the Mercy Hospital for Women in Melbourne, located 12km from RCH. Contact with RCH was made three weeks from the planned delivery, requesting anaesthetic and surgical support for the neonate. A team consisting of two specialist Paediatric Anaesthetists, an ENT surgeon, Paediatric surgeon, scrub/scout nurses and an Anaesthesia Technologist were assembled.

The mother was scheduled for a Caesarean section under general anaesthesia. A lower abdominal skin incision (plannenstiel incision) was performed then optimal neonatal position was identified using ultrasound on the uterus prior to uterine incision. Following the incision the neonate’s head and shoulder was delivered and the sterile neonatal anaesthetic team begins to establish the neonate’s airway (Costello et al., 2010).

Attempts to secure the airway were to be made in the following order:
- direct laryngoscopy with traditional intubation
- flexible fibre optic laryngoscopy
- rigid bronchoscopy to be performed by the ENT team
- tracheostomy - until an adequate airway was secured
The MWH is an obstetric hospital primarily focussed on the treatment of adult patients, however was provisioned for the resuscitation of neonates and has a NICU onsite. However after discussion between the Anaesthetists and myself, the Anaesthesia Technologist, a decision was made to take the following equipment so that familiar equipment was on hand for the Paediatric Anaesthesia team at the delivery.

The equipment taken was:

<table>
<thead>
<tr>
<th>Equipment</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glidescope screen, plus paediatric baton and size zero and size one glide scope blades and small glide scope stylets</td>
<td>Laryngoscope handle small thick and stubby in height</td>
</tr>
<tr>
<td>Size 2.8 fibre-optic bronchoscope with C-Mac attachment cable allowing the fibre optic picture to be shown on the C-Mac screen to allow portability rather than larger tower</td>
<td>Laryngoscope handle long and thin</td>
</tr>
<tr>
<td>Uncuffed endotracheal tubes sizes 2.0, 2.5, 3.0 and 3.5 x3</td>
<td>New Batteries in each and spare</td>
</tr>
<tr>
<td>Elastic rope intubating boujie</td>
<td>Mackintosh blades 0,1</td>
</tr>
<tr>
<td>Small stiff intubating boujie</td>
<td>Riester blades 0, 1</td>
</tr>
<tr>
<td>ETT exchange catheter with removable inner stylet</td>
<td>Miller blades 0,1 in both disposable</td>
</tr>
<tr>
<td>LMAs size 1 &amp; 1.5</td>
<td>Seward blade 1</td>
</tr>
<tr>
<td>Orophyngeal airways sizes 0,00</td>
<td>Rigid suction Yanker handles small and large</td>
</tr>
<tr>
<td>Nasal phyngeal airways 2.5, 3.0 and 3.5</td>
<td>Soft suction catheters size 6, 7 and 8 Fr.</td>
</tr>
<tr>
<td>Two anaesthetic face masks with filters and elbows – extra small and small</td>
<td>Ayers T-piece with a Jackson Rees modification</td>
</tr>
</tbody>
</table>

This equipment was packed, in preparedness for immediate transport- in case of an emergency delivery – such as if the mother went into early labour.
On the day of the procedure the RCH team travelled to the MHW by taxis.

At the MHW, a large-scale briefing of all the involved health professionals was undertaken, including an orientation of the theatre complex. Due to the large size of the team staff were identified by both nametags as well as coloured theatre caps identifying their role in theatre. Additionally the paediatric anaesthesia and surgical teams also conducted briefings focussed on the management of the neonate once partially delivered.

Because of the nature of the EXIT procedure, the neonate airway is secured while the baby was still partially delivered by Caesarean section – occurring in the sterile field of the mother.

In effect, the placental circulation acts as a naturally occurring Extra Corporeal Membrane Oxygenation (ECMO) circuit.
Photo 4 & 5: Sterilized Anaesthetic Equipment
Consequently the airway equipment had to be sterilised and anaesthetic personnel wear sterile gowns and gloves.

The mother was anaesthetised for general anaesthetic caesarean section; large bore intravenous cannulas placed as well as intra-arterial cannulas. The mother was positioned in low lithotomy with both arms out at 90 degrees on arm boards.

The neonate was then identified on ultrasound and a needle guided into the neonate’s neck mask in an attempt to drain some of the mass to reduce its size and make the delivery and intubation easier. 90mls of serosanguinous fluid was drained percutaneously prior to commencing the Caesarean. The neonate’s head and right shoulder was then delivered and the anaesthetic team moved into place, a foetal heart monitoring scalp clip was connected to the shoulder.

The primary anaesthetist attempted to palpate the tracheal location externally but discovered this was largely obscured by the neck mass. Intubation was attempted with a Mackintosh Zero blade, with the anaesthetist discovering significant airway oedema distorting and obscuring the airway anatomy and location of the laryngeal inlet. An attempt was made to place a small rigid bougie into what appeared most likely to be the larynx, with a size three uncuffed endotracheal tube inserted over the bougie. After placement of the endotracheal tube clinical confirmation was made by connecting a T-piece and observing chest movements on the still partially delivered neonate. The supporting anaesthetist then confirmed anatomical placement of the ETT using a 2.8mm fibre optic bronchoscope, with confirmation by observation of the tracheal rings. Only after the ETT was confirmed and secured properly was the placental circulation severed and the neonate handed over to the care of the Mercy Hospital neonatologists. The neonate was further resuscitated in theatre on an Ohio neonatal resucitaire prior to transfer to neonatal intensive care unit (NICU).

Once stabilised at the Mercy Hospital, the neonate was transferred to the RCH NICU for treatment of their condition. The neonate was unsuitable for extubation and had a surgical tracheostomy performed. The lymphatic malformation was treated by surgical dissection, and sclerotherapy and the neonate continues to be managed by multiple teams at the Royal Children’s Hospital.

**Practical Challenges During the EXIT Procedure**

Logistically, making room in the theatre set up for the sterile anaesthetic technologist to be on the anaesthetists right, due to placement of the
ultrasound and neonatologist this was not considered and made passing equipment across the anaesthetist body difficult. Unfortunately the scalp clip was dislodged, and pulse oximetry readings could not be reliably made due to the amniotic fluid on the neonate until they were dried on the Ohio.

If the intubation process persisted a time taken to put a new scalp clip of dry the neonates hand to enable monitoring. During the intubation the ultrasound was used to monitor the neonates heart rate.

Having a sterile T-piece is for future use, although the filter, mask and tubing was sterile the delivery bag was not and the second anaesthetist had to desterilise to use the bag. Getting a sterile T-piece set up put together and processed prior to the event, as a means of delivering ventilation was important in establishing correct airway placement (Hirose & Harrison, 2003).

Other difficulties identified were that of unfamiliar environment, unknown staff and some distance away from the anaesthetics team base hospital. The essentials of the difficult intubation trolley, which is in use at the base hospital had to be packed up into medical suitcases and set up on small metal trolleys. In order to prevent any further problems logistics and equipment, two or three of everything was brought were appropriate. For example endotracheal tubes, laryngoscope blades, tube connectors, bougies masks oropharyngeal airways.

**Discussion of the EXIT Procedure**

The exit procedure allows partial delivery of the foetus with prenatally diagnosed airway obstruction allowing management of the compromised airway while the neonate can rely on continual placental function (DeValle et al., 2014). Clearly as the preceding case illustrates significant planning and preparation is required to successfully undertake an EXIT procedure. Multiple teams are required, and good communication between teams is essential (Abbie, 2011).

The EXIT procedure takes advantage of an opportunity where a neonate can be partially delivered by caesarean section, and have otherwise lethal airway issue addressed while still perfused by the placental circulation (Hartnik et al., 2009).
Only after the airway is satisfactorily secured is the umbilical cord clamped and cut and the neonate becomes reliant on its own airway (Iserte et al., 2012).

**Indications for the EXIT Procedure**

The primary causes for prenatally diagnosed upper airway obstructions are that of head and neck teratomas, lymphatic/vascular malformations, severe micrognathia and congenital high airway obstruction syndrome (Mong et al., 2008).

**TERATOMAS**
Teratomas are germ cell tumour with independent cell growth and the potential to change normal cell tissue next to them. They are often not malignant but can extend from mandible to clavicle and include everything from the tongue to the roof of the mouth. These facial tumours distort the airway with their solid uncompressible size making direct laryngoscopy difficult (Saito-Benz et al, 2017).

**LYMPHATIC/VASCULAR ABNORMALITIES**
Lymphatic/Vascular abnormalities are the most common reason for exit procedure as they occur most frequently in the neck and head. Lymphatic
Malformations are benign and come from embryological disturbances in the development of the lymphatic system. These malformations are infiltrative rather than solid and can be drained prior to the procedure to reduce some of the size and allow easier airway attempt (Hartnik et al., 2009).

**SEVERE MICROGNATHIA**
Severe micrognathia is the underdevelopment and retro displacement of the mandible. This can displace the tongue base causing complete airway obstruction, only severe cases require airway establishment during an exit procedure. Severe cases are often very difficult to intubate with laryngoscopy and need either fibre optic intubation or tracheostomy (DeValle et al, 2014).

**CONGENITAL HIGH AIRWAY OBSTRUCTION SYNDROME**
Congenital high airway obstruction syndrome (CHAOS) includes a broad group of airway disease from laryngeal web to complete laryngeal or tracheal stenosis. The characteristics of these abnormalities are identified by the ultrasonography findings of dilated airways distal to the obstruction, large hyper echogenic lungs with diaphragmatic inversion. Can also present with such features as subcutaneous oedema, ascites and polyhydraminos. CHAOS was previously fatal, the exit procedure has ensures the controlled delivery of the effected neonate and airway establishment by often a tracheostomy (Mong et al., 2008).

Often these Airway abnormalities are associated with a congenital syndrome, including a chromosomal abnormality, which requires additional paediatric management. The presence of such abnormality increases overall mortality (Bourchard et al., 2002).

**Contraindications for the EXIT Procedure**

Contraindications to perform an EXIT procedure include a significant disabling or non-life sustaining structural abnormality of the foetus and genetic abnormality, such that the procedure would be futile or lead to an extremely poor quality of life.

The welfare of the mother must also be considered.

Potential additional risks to the mother, from having the EXIT procedure include experiencing significant blood loss from uterine hypotonia, increased risk of postoperative wound infections from the increase of personnel in the operating theatre, and the additional duration of the procedure (Flipchuk & Avdimirentz, 2009).

Risks to the neonate include, inability to establish an airway leading to hypoxia and brain damage and hypothermia. The neonatal body temperature could drop dramatically due to the exposure, combined with immaturity with reduced body fat to provide insulation (Mayer et al., 2010).
Conclusions

The EXIT procedure was used successfully to ensure uteroplacental gas exchange and the foetus remained stable during the securement of a difficult airway. Prior to the advent of EXIT procedures neonatal mortality associated with these conditions was as high as 80% (Bourchard et al., 2002). A well planned, organized and controlled procedure in which one is prepared to face the most difficult situations will maximise the chance of success.

Photo 6: Theatre Doors at MWH
References


