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REVIEW

Fetal airway management on placental support: Limitations and ethical considerations in seven cases

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The aim of this study was to evaluate the paediatric and maternal outcome after *ex utero* intrapartum treatment (EXIT). A retrospective review was carried out of the medical charts (gestational age, circumstances of diagnosis, multidisciplinary prenatal decision, date of surgery, paediatric and maternal outcome) of all the fetuses eligible for/delivered via the EXIT procedure in our paediatrics and obstetrics tertiary care and teaching centre, between October 2004 and May 2011. Seven fetuses with cervical teratoma, epignathus tumour or congenital high airway obstruction syndrome (CHAOS) were included in our study. Two pregnancies were terminated and five fetuses were delivered alive. The airway was secured in all five cases (two endotracheal intubations and three tracheostomies). No maternal complications were observed. On average, babies were delivered at 32 gestational weeks, and spent 31 days in the intensive care unit. All but one baby were ventilated for 18 days. Long-term paediatric outcome was favourable. It is concluded that airway management by the EXIT procedure has become an efficient technique. A multidisciplinary prenatal assessment is essential in order to select appropriate cases.

Keywords: Cervical teratoma, CHAOS, epignathus, EXIT, fetal surgery, placental support

Introduction

Fetal airway obstruction at birth can result in profound hypoxia, anoxic brain injuries or even death. The main aetiologies of congenital fetal airway obstruction are cervical tumours, such as teratomas or lymphangiomas, congenital laryngeal stenosis or atresia as in congenital high airway obstruction syndrome (CHAOS) and iatrogenic tracheal obstruction by balloons in cases of severe congenital diaphragmatic hernia. Advances in prenatal ultrasound and magnetic resonance imaging (MRI) techniques have led to improvements in the management of airway obstruction (Hirose et al. 2004). Operations on placental support (OOPS), also referred to as *ex utero* intrapartum treatment (EXIT), are performed before cutting the umbilical cord and by maintaining delivery of oxygen to the fetus through the placental circulation, thus allowing fetal airway management by intubation or tracheotomy.

The first successful case was described in 1992 by Catalano and co-workers, in a fetus presenting with an epignathus tumour. Since then, several cases of cervical tumours such as teratomas leading to airway compression have been successfully managed, with excellent functional and cosmetic outcome (Azizkhan et al. 1995).

The EXIT procedure is also promoted in instances of fetal endoluminal tracheal occlusions (FETO) for severe congenital diaphragmatic hernia in order to recover fetal airway patency at birth. In cases of premature delivery, the EXIT procedure permits the surgeon to remove the tracheal balloon without anoxic sequelae.

Another indication is CHAOS, which groups together a spectrum of anomalies, including laryngeal web, laryngeal atresia, laryngeal cyst and tracheal atresia or stenosis, with complete or near complete obstruction of the airway. While CHAOS was initially considered as incompatible with survival, the development of placental support techniques and performance of early tracheotomy in the delivery room have led to reports of several cases with good mid- and long-term outcomes (Crombleholme et al. 2000; Hirose and Harrison et al. 2003; Shimabukuro et al. 2007).

The main challenges facing these procedures are: (1) determining the aetiology and the degree of fetal airway obstruction, and (2) combining airway management on placental support/EXIT with minimal maternal morbidity and good fetal outcome.

Careful management of the umbilical cord is not sufficient to ensure correct fetal oxygenation. An anaesthetic protocol promoting complete uterine relaxation and avoiding any decrease in feto–placental flow is crucial. However, such relaxation also increases the possibility of uterine haemorrhage (Butwick et al. 2009; Skarsgard et al. 1996). A fine balance is therefore required and strict protocols have now been established, which have allowed successful interventions without significant maternal morbidity.

The purpose of this paper is to evaluate fetal and maternal outcomes of this procedure based on our experience and reports in the literature and to establish guidelines concerning its indications and limitations.

Materials and methods

The study required a retrospective review of our institutional and departmental database and was approved by the local Institutional Review Board. Included in the study were all fetuses eligible

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for/delivered via the EXIT procedure between October 2004 and May 2011, with a medical chart containing information on initial diagnosis, prenatal imaging, operative reports, feto-pathology reports and outpatient outcome.

Protocol in our centre

Decision-making process regarding airway management on placental support via the EXIT procedure

In France, three prenatal ultrasounds for the detection of fetal anomalies are recommended at 12, 22, and 32 gestational weeks. Some reveal an airway obstruction by such, as a cervical or facial tumour, hydramnios (secondary to interference with fetal swallowing), large echogenic lungs, flattened or everted diaphragms or dilated distal airway (secondary to CHAOS). In the latter case, fetal and maternal size and tolerance are regularly assessed by ultrasound.

All women included in our series underwent prenatal ultrasound at the recommended stages of pregnancy. A complementary fetal MRI then provided more detail concerning the size, position and anatomical relationship of the airway compression suspected on ultrasound, and contributed to exploring the boundaries and content of cervical tumours (Coakley et al. 2004). Based on the imaging data, a multidisciplinary team, including obstetricians, neonatologists, anaesthetists, paediatric ENT surgeons and radiologists, evaluated any indication of a need for airway management on placental support. The risks and potential benefits were then explained to the parents and family consent was obtained.

All cases were considered as at-risk of prematurity, and pulmonary maturation was induced by intramuscular steroids; two successive daily injections (betamethasone or dexamethasone, 12 mg) whenever possible.

Anaesthesia and airway management during the procedure

The procedure used is based on the principles of fetal surgery under general materno–fetal anaesthesia. Uterine relaxation was achieved using a high concentration of halogenated inhaled anaesthetic agents (2–4 minimal alveolar concentration). Transplacental passage of maternally inhaled anaesthetic agents resulted in fetal anaesthesia 10–15 min after achieving maternal anaesthesia. Maternal blood pressure was monitored and maintained high with ephedrine if necessary to ensure placental perfusion. Atropine was usually administered. A low transverse incision of the abdominal skin and hysterotomy was then performed. The aim was to keep fetal exposure to a minimum to reduce heat loss, preserve uterine volume and reduce the risk of umbilical cord compromise or premature placental detachment (Marwan and Crombleholme 2006). Such manoeuvres are intended to preserve uteroplacental circulation and a continued gas exchange. When full exposure of the fetus was required to facilitate airway control (Preciado et al. 2004), the fetus was placed in a sterile bag to minimise heat loss and in a position lower than placenta level, taking care to avoid any tension on the umbilical cord. When necessary, direct fetal anaesthesia was performed by intramuscular injection, to avoid cardiac distress. Fetal haemodynamics were monitored using a pulse-oximeter and, whenever possible, a heart-rate monitor or echocardiography. If the uterus was too tense, sublingual glycerol trinitrate was administrated to the mother before the cord was clamped.

Direct laryngoscopy using rigid 2.7 mm 0° rigid telescopes (Karl Storz, Tuttlinger, Germany) was primarily performed in order to evaluate the laryngeal aspect before endotracheal intubation was then attempted. When intubation was impossible, bronchoscopy or tracheotomy was performed, based on a pre-established algorithm. Once the airway was secured, and before clamping the umbilical cord, coordination between surgeons and anaesthetists prevented uterine atony and excessive maternal bleeding; inhaled agents were decreased and intravenous oxytocin was delivered to the mother. If needed, sulprostone was administered to reinforce uterine tone and reduce the risk of bleeding. Multiple blood vessel accesses were ensured and blood products were reserved in advance. The EXIT procedure was carried out as quickly and efficiently as possible in all cases.

Results

Our series included five babies born after airway management on placental support, via an EXIT procedure (four cervicofacial teratomas and one CHAOS) and two cases of medical terminations of pregnancy (two teratomas) between October 2004 and May 2011.

Babies born after EXIT procedure

Case 1

A 5 cm diameter left-sided cervical mass was discovered by ultrasound at 30 weeks' gestation. Fetal MRI showed an absence of liquid in the tracheal lumen due to tumoural compression (Figures 1 and 2). The mother was hospitalised at 32 weeks for premature labour unresponsive to pharmacological treatments, and intravenous steroids were started, to induce pulmonary maturation. After parental consent, the mother underwent an emergency EXIT procedure under general anaesthesia, followed by a caesarean section. Direct laryngoscopy showed a complete subglottic obstruction; endotracheal intubation and bronchoscopy were initially unsuccessful. The fetus was therefore tracheotomised before the cord was clamped. Once delivered, the baby's ventilation became unstable; another bronchoscopy

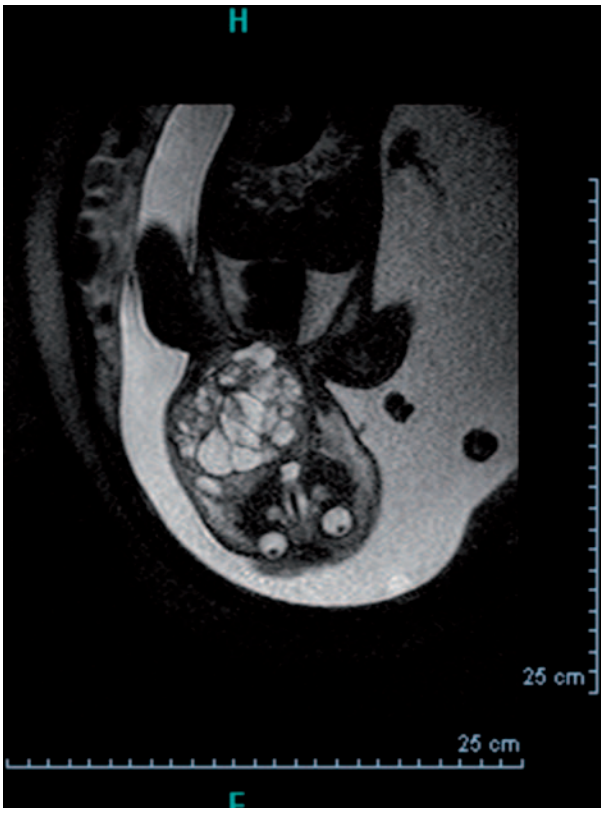

Figure 1. Frontal fetal MRI at 30 gestational weeks, showing tracheal compression by a cervical mass.

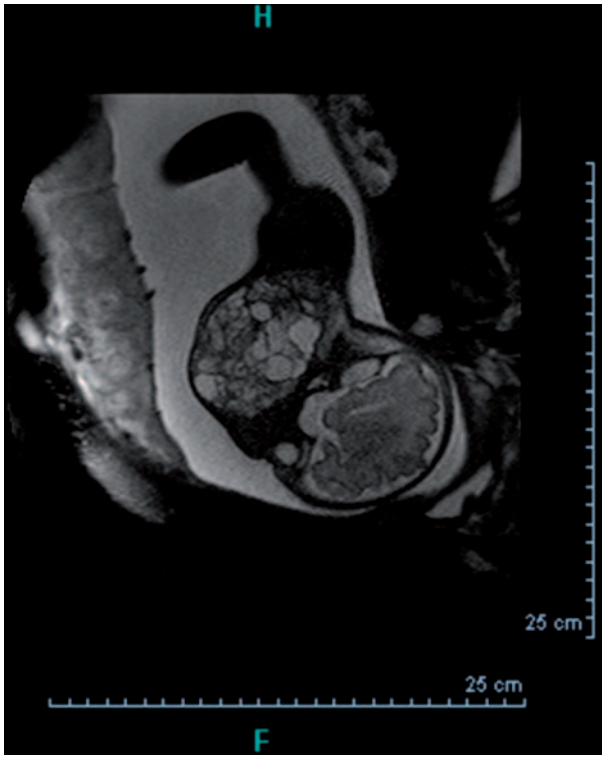

Figure 2. Sagittal fetal MRI at 30 gestational weeks, showing tracheal compression by a cervical mass.

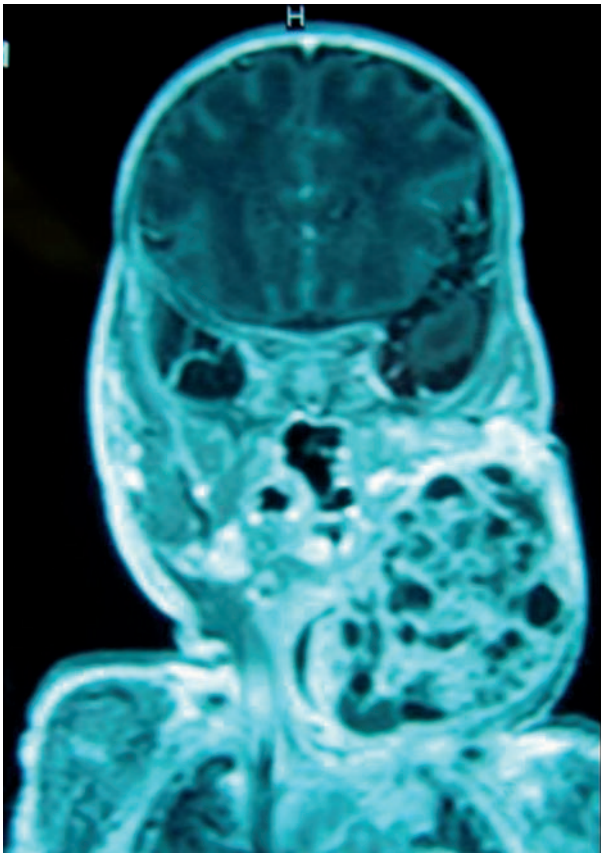

Figure 3. Preoperative MRI of the first baby with cervical teratoma.

was performed, and this time gave access to the tracheal lumen, with the help of external movements of the mass and retrograde laryngotracheal catheterisation through the tracheal opening. A naso–tracheal tube was then placed and the tracheal incision was sutured. A pneumothorax was diagnosed and exsufflated without complication. The total period on placental support was 20 min. A postnatal MRI was performed in order to assess the cervical teratoma (Figures 3 and 4). The tumour was surgically excised at Day 5; it originated from the left thyroid and no left laryngeal nerve could be identified (Figures 5 and 6). A postoperative endoscopy confirmed left vocal cord paralysis. The baby was extubated at Day 11. Two months later, a right thyroid mass was diagnosed along with an increase in alpha-fetoprotein. A revision surgery was performed and the contralateral thyroid lobe was removed. The baby was discharged 3 months later. During 7 years of follow-up, the child has shown no recurrence and is doing well.

Case 2

A very large anterior teratoma of the neck originating from the left thyroid was detected during pregnancy on routine ultrasound, at 27 gestational weeks. The fetal MRI confirmed an 8 cm diameter cervical heterogeneous mass, with complete laryngotracheal compression. Pulmonary maturation was induced prior to total delivery of the baby by caesarean section at 33 weeks, with preservation of the umbilical cord. Intubation was obtained with a size 2.5 bronchoscope, which was immediately replaced with a 3 mm inner diameter (ID) orotracheal tube. The period of time on placental support was 5 min. Bulging of the teratoma into the pharyngeal region prevented a more stable nasotracheal intubation and immediate surgery was performed after postnatal MRI. The laryngotracheal axis was embedded within the tumour and the cricoid cartilage appeared thin and distorted, particularly on the left-hand side, where the recurrent laryngeal nerve could not be identified. Postoperatively, several complications (left laryngeal immobility, immature lungs, glottic oedema) rendered extubation impossible and the baby was tracheotomised after 1 month of intubation. Laryngotracheal reconstruction and segmental tracheal resection were then required before decannulation. He is now 6 years old and doing well.

Case 3

CHAOS syndrome, due to complete laryngeal obstruction was suspected at 28 gestational weeks, after identification on routine ultrasound during pregnancy of large echogenic lungs and a flattened diaphragm. A fetal MRI at 30 gestational weeks confirmed the diagnosis of complete laryngeal obstruction (Figure 7). At 31 weeks, after failure of tocolysis, a rapid vaginal breech extraction

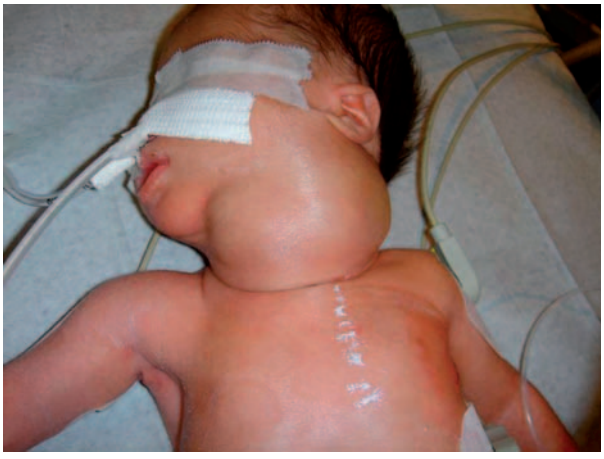

Figure 4. Preoperative view of the first baby with cervical teratoma.

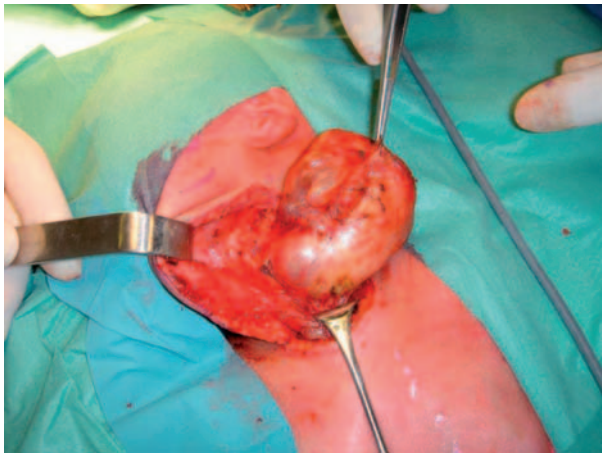

Figure 5. Intraoperative view of the first baby.

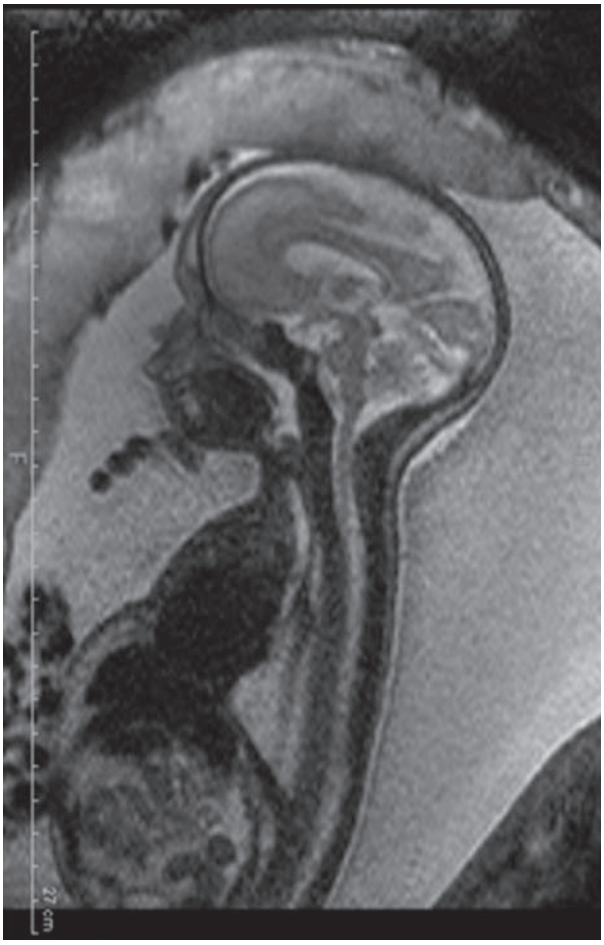

Figure 7. MRI of the CHAOS, with a sign of obstruction being the disappearance of the liquid signal from the trachea.

with umbilical cord maintenance was performed under general anaesthesia, following the mother's refusal of a caesarean section. Direct laryngoscopy confirmed complete obstruction of the subglottis by a thick membrane, and a tracheotomy was immediately performed. Once the trachea was opened, a large amount of clear pulmonary fluid was sucked out and the cord was subsequently clamped. The procedure lasted 20 min.

The baby later showed signs of chronic respiratory distress due to immaturity of abdominal and diaphragm muscles and motor retardation. He underwent a gastrostomy and was mechanically ventilated via the tracheotomy. As an infant, he walked at 21 months. He is now 6 years old, has near-normal oral feeding, shows no sign of mental retardation and has been weaned from nocturnal ventilation. Prematurity and/or CHAOS may be partly responsible for these earlier complications. A laryngotracheal

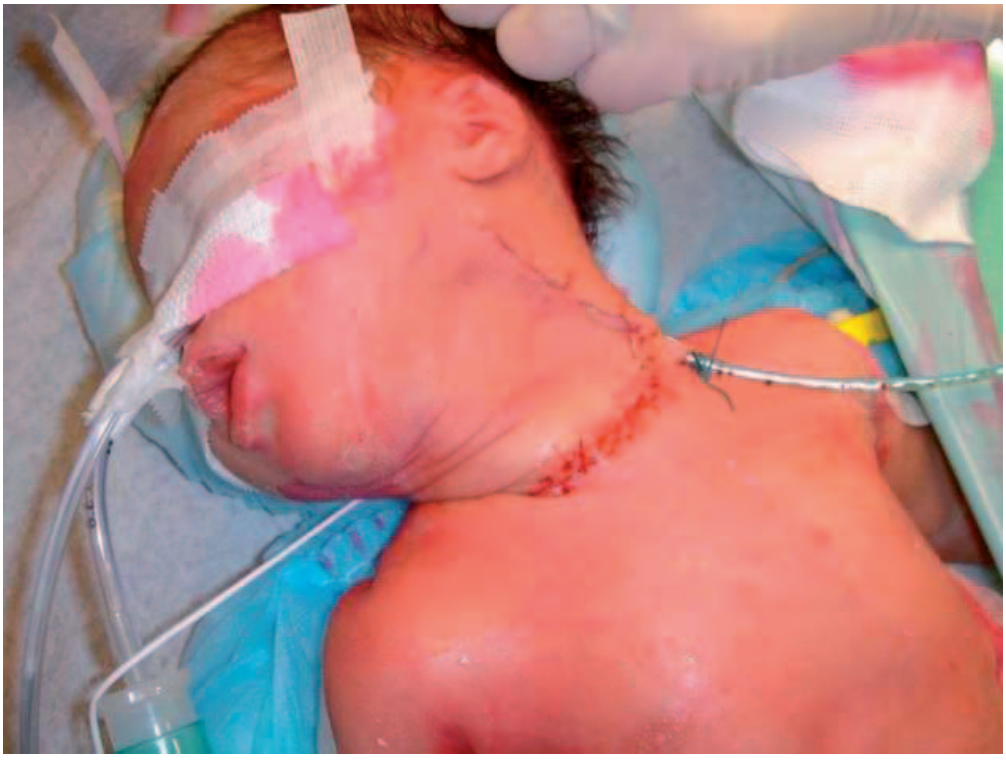

Figure 6. Postoperative view of the first baby.

reconstruction has now been performed allowing a successful decannulation.

Case 4

An epignathus tumour was suspected in the fetus at 24 gestational weeks, on routine ultrasound during pregnancy. The fetal MRI confirmed complete nasopharyngeal obstruction by a lesion protruding through the left nasal cavity and extending to the laryngeal inlet. Caesarean delivery was performed at 33 weeks, under general anaesthesia. Direct laryngoscopy showed no infiltration of the laryngeal inlet, and orotracheal intubation was successful. However, due to concerns over intubation instability, surgical excision of the lesion was performed within the first few hours of delivery. Local recurrence of a choanal tumour bud required surgery at 5 months. Other than this, the postoperative follow-up has been uneventful and at 3.5 years of age, the child is doing well.

Case 5

The third routine ultrasound during pregnancy revealed a voluminous nasal and oropharyngeal mass. Fetal MRI at 31 gestational weeks confirmed a large epignathus tumour. Considerable hydramnios, twice requiring evacuation via amniotic puncture, as well as rapid growth of the mass, led to a caesarean section being performed at 34 weeks, after pulmonary maturation was confirmed.

Orotracheal intubation was impossible by direct laryngoscopy; a tracheotomy was therefore performed. The cord was clamped just after tracheotomy, and the baby girl was immediately transferred to the operating room for tumour resection.

Surgical excision was achieved after section of the central part of the velum. Conventional nasopharyngeal intubation was then performed before closure of the tracheotomy.

During the follow-up, she showed signs of pulmonary immaturity despite corticosteroids and surfactant. She was discharged at 40 days and at the age of 6 months, she is doing fine.

Results

On average, these babies were born at 32 weeks and spent 31 days in the intensive care unit. Except for Case 3, who was ventilated for a few hours daily until the age of 5 years, the mean duration of mechanical ventilation was 18 days.

The outcome was favourable for all mothers: they were discharged after 3–7 days (mean 4.3 days). None suffered excessive postpartum haemorrhage.

Terminated pregnancies

During the study period, two other fetuses were diagnosed as having teratomas serious enough to motivate a medical termination of the pregnancy, after multidisciplinary discussion and parental consent. In one case, the MRI suggested a teratoma originating at the base of the tongue and completely filling the mouth and the laryngeal aperture. Surgical excision with removal of the base of the tongue would have led to permanent and severe swallowing disorders. With regard to the other case, an MRI at 27 gestational weeks showed severe oedema and ascites of the fetus, and a pulmonary hypoplasia leading to a poor prognosis associated with abnormal vital functions. In both cases, the parents accepted the termination; the procedure was performed under epidural anaesthesia for one and general anaesthesia for the other. Follow-up was uneventful. Feto-pathological analysis confirmed the poor prognosis in the second case, however it revealed a limited implantation in the nasopharynx for the first in contrast to the more serious implantation in the tongue suggested by the MRI.

Discussion

In our series of patients, the medical decision to perform an EXIT procedure was, in all cases, approved by both parents. Consent was obtained following a multidisciplinary consultation with obstetricians, an ENT surgeon, anaesthetists and a neonatologist, during which the procedure, including associated risks and benefits, were explained, the parent's questions answered and psychological support proposed.

Delivery conditions for airway management on placental support

Premature delivery is frequent among this patient population and may be due to hydramnios caused by fetal swallowing disorders. All the babies in our series were delivered prematurely. To reduce the risks associated with hydramnios, obstetricians prenatally performed evacuation punctures and also induced pulmonary maturation with corticosteroids.

The role of the ENT surgeon is important during these deliveries in obtaining and maintaining the newborn's airway (Figure 8). We recommend thorough preparation with reference to a protocol checklist (see Table I) to ensure that all the material needed for laryngoscopy, bronchoscopy and tracheotomy is available in the delivery room. The team's experience is crucial for success.

In our opinion, the high risk of acute respiratory obstruction in cases of accidental extubation justifies nasotracheal intubation wherever possible. This corresponds to the most stable choice in neonates, however it may be more difficult to perform than an orotracheal intubation. For this reason, unlike other authors (Hedrick et al. 2003), we preferred to deliver these babies as completely as allowed by the length of the cord without any tension (Figure 9). The desired outcome of the EXIT procedure is delivery. Maximum comfort and good access to the airway are essential for success, particularly in cases of large cervical tumours. They also minimise the duration of the procedure, which helps reduce the risk of maternal haemorrhage, fetal hypothermia or anoxia. This differs from intrauterine surgery, after which the pregnancy continues for several weeks, and during which, any cord tension can induce premature labour or *in utero* death (Kohl et al. 2006).

The mean duration of materno–fetal circulation is about 20 min, even without anaesthesia management. Some have extended this time to 2.5 h to allow for a complete surgical procedure with no negative consequences for the baby or the mother; however, the maternal risk is greater during such long procedures (Butwick 2009; Hirose et al. 2003; Noah et al. 2002).

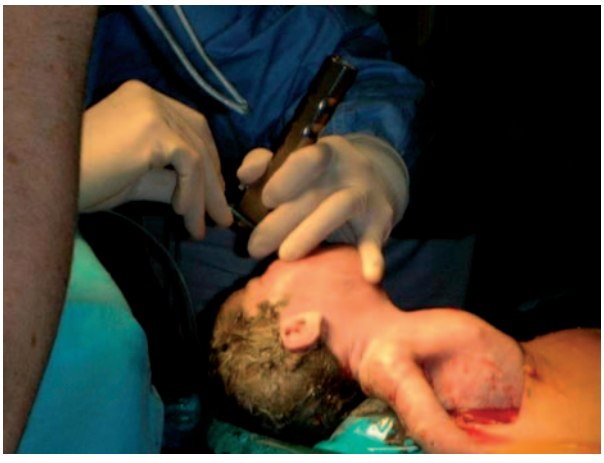

Figure 8. Laryngoscopy on placental support.

Table I. Checklist protocol for the EXIT procedure.

	Check
With sterile presentation	
A set of laryngoscopes fitted for the weight of the baby (frequent prematurity)	
A set of intubation tubes of 2, 2.5, 3	
A MAGILL Tweezers	
A set of mounted rigid bronchoscopes in sizes 2, 2.5, 3	
A power cable for cold light	
A tracheotomy set	
A set of tracheotomy cannulae in neo-sizes 2.5–3.5	
A set of sterile and warm sheets , baby bag	
Baby monitoring equipment	
A small instrumental table (to settle the baby between the mother's leg)	
A reflectance pulse oximeter	
Eventually an EKG scope	
Intravenous catheters in case of vital distress (24 G)	
Oxygen source and mask for ventilation	

Among our series, the EXIT procedure lasted at most 20 min, during which time two fetuses were quickly intubated and three needed a tracheotomy.

In one case in our series (Case 3, CHAOS syndrome), the mother refused a caesarean section. The EXIT procedure was performed following vaginal delivery, which appeared to be the best compromise between maternal and fetal risks. The risk of dystocia was low due to the small weight of the baby and the absence of a cervical mass. To our knowledge, this is the first case described in the literature. This baby required a tracheotomy that was performed without difficulty. The outcome for the mother was uneventful.

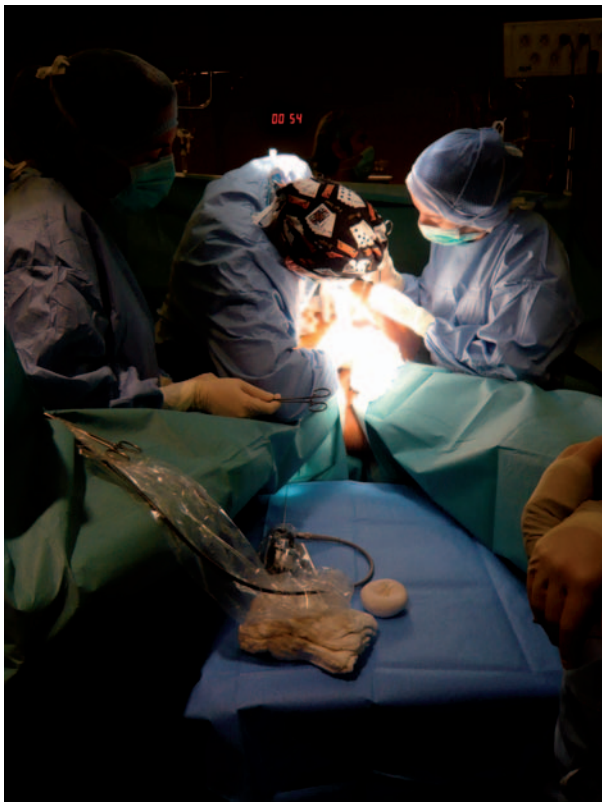

Figure 9. Installation during the EXIT procedure; the table between the mother's legs is for the baby in anticipation of delivery.

Planning surgical treatment

Early post-EXIT procedure events should always be anticipated. In cases where the management of the airway is considered highly stable (nasotracheal intubation, removal of tracheal balloon, tracheotomy in babies over 3 kg), corrective surgery can be delayed. However, following orotracheal intubation or tracheotomy in babies weighing < 3 kg, frequently associated with cervical or pharyngeal mass, surgery should be performed within a few hours once the newborn has been stabilised by the neonatologist based on the results of selective imaging studies (Figures 10 and 11). Some authors have reported removal of teratomas or cervical tumours under placental support during the EXIT procedure (Hirose and Sydorak et al. 2003). Among our series of patients, a short EXIT procedure limited to securing the airway of the fetus followed by early surgery appeared preferable to complete excision surgery under placental support (Noah et al. 2002; Preciado et al. 2004). In Cases 1 and 2, the preservation of important vital organs (carotid and jugular vessels, trachea) and functional structures (cranial nerves) was difficult due to modification of anatomical landmarks, and careful dissection was required (Sichel et al. 2002).

Ethical considerations

The new possibilities for management of such high-risk newborns sometimes introduce an ethical dilemma between saving the life of the baby and protecting that of the mother. Parents must be well informed of both the fetal and the maternal events relating to the procedure and the unpredictable evolution of each child's status, depending on the aetiology of the airway obstruction.

Before the development of the placental support/EXIT procedure, such neonates had a life-threatening and/or poor neurological prognosis, due to frequent sequelae of anoxia or prolonged hypoxia (two out of 17 patients in the series of Azizkhan et al. 1995). Complications still exist even under placental support, although they are difficult to quantify, since it is unclear whether they are related to the aetiology or to the procedure itself.

The global prognosis does however, seem highly dependent on aetiology, and on prematurity, which is frequent in these babies (all of the cases in our series). Respiratory distress due to pulmonary immaturity is frequent despite prenatal use of corticosteroids. For example, our third case (CHAOS syndrome) was still tracheotomised and dependent on mechanical ventilation after 4 years due to immaturity of the respiratory muscle and diaphragmatic dysfunction, which consequently delayed laryngotracheal reconstruction.

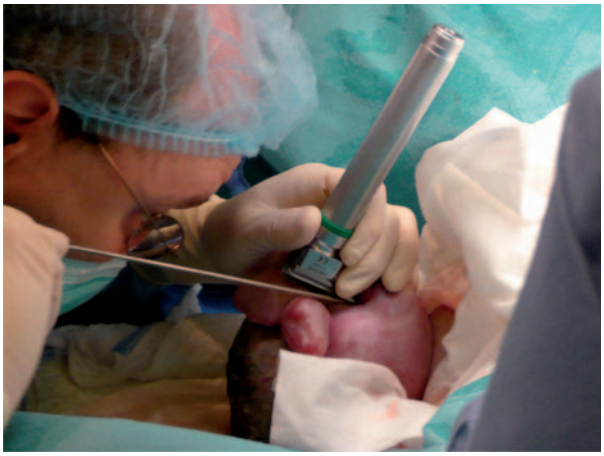

Figure 10. Direct laryngoscopy attempting to expose the larynx.

Figure 11. Following tracheotomy, the child was transferred to the operating room for immediate surgery.

While the delivery of fetuses presenting with teratomas is difficult, their long-term survival is excellent (84%). Prior to the placental support/EXIT procedure, the mortality rate of such babies was 43% in those showing respiratory distress at birth and 27% in those not, as reported by Jordan and Gauderer (1988). The two babies in our series with cervical teratoma had subsequent paralysis of the left vocal cord relating to the thyroid/cricoid insertion of the tumour and probable disturbed growth of the laryngeal recurrent nerves. One required several open surgical procedures before decannulation was possible. In 2002, Sichel and co-workers reported a similar case and concluded a probable agenesis of the recurrent laryngeal nerve as well as an anomaly of the carotid artery, internal jugular vein, and thyroid gland.

Prognosis seems less favourable in cases of congenital diaphragmatic hernia, for whom the survival rate is 38.5%, due to comorbidities, such as pulmonary hypoplasia (Hedrick et al. 2003).

Particularities of epignathus teratomas

One of the fetuses that was medically aborted in our series revealed discrepancies between the MRI aspects and feto-pathological findings. Clinicians should be aware that prenatal MRI cannot always distinguish the insertion site of the teratoma. Most reported cases of epignathus teratomas originate from the skull base (sphenoid region), the pharynx (Rathke's pouch) or the hard palate. No instance of insertion on the tongue base has previously been described.

In addition, intracranial extension is a key prognosis factor for this pathology, and encephalic structures must be carefully screened on the MRI (Calda et al. 2011; Tonni et al. 2010).

Maternal risks

In a retrospective study comparing EXIT and caesarean section procedures, Noah et al. (2002) found that the duration of the EXIT procedure is twice that of caesarean section, and that postpartum haemorrhage was more likely to occur during the EXIT procedure. However, no differences were found in postoperative haematocrit level or postpartum length of stay. Women undergoing the EXIT procedure had more wound complications but the number of cases of endometritis was no different (Noah et al. 2002). The EXIT patients were treated more frequently with antibiotics, though the small difference between the two groups was probably due to a closer surveillance of those undergoing the EXIT procedure. Another team has reported a severe haemorrhage consecutive to an EXIT procedure (Butwick et al. 2009). The use of uterotonic drugs, a strict adherence to anaesthetic protocol, and perfect coordination between obstetricians and ENT surgeons to minimise the duration of the procedure are essential to prevent excessive maternal bleeding.

Conclusion

Airway management on placental support during the EXIT procedure has improved the prognosis of fetuses presenting with a life-threatening airway obstruction at birth. In our experience, the maternal morbidity and mortality rates are very low and justify using this approach.

Effective management requires pre-, peri- and postnatal input of a multidisciplinary team composed of radiologists, obstetricians, neonatologists, anaesthesists and paediatric ENT surgeons. Multidisciplinary prenatal decision meetings are taken to assess the risk of airway obstruction as well as the short- and long-term fetal prognosis; the role of the different participants in the delivery room should also be made clear and a management strategy planned. The specific role of the ENT surgeons is to secure the airway (intubation, tracheotomy, etc.) in the delivery room as quickly and as safely as possible, to stabilise the fetus, and to organise the postnatal surgery of the newborn, which may be required as early as within the first few hours of birth. This requires an organised and experienced team of neonatologists and obstetricians.

Information given to the parents about immediate consequences, long-term prognosis and unpredictable complications must be as complete as possible. Long-term follow-up of these children is also important, due to the risk of recurrence and/or sequelae (due to either pathology or prematurity) of, e.g. vocal cord paralysis or tracheomalacia.

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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