CASE REPORT

Ex utero intrapartum treatment (EXIT) of severe fetal hydrothorax

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Ex utero intrapartum treatment (EXIT) of a fetus with severe bilateral hydrothorax is described. EXIT allows therapeutic interventions on the neonate while maintaining fetoplacental circulation. Thus it may be useful for fetuses presenting with severe pleural effusion towards the end of gestation and in whom in utero drainage is not possible or available and drainage post partum would result in profound and prolonged hypoxia until sufficient drainage of pleural fluid allowed lung expansion.

Congenital hydrothorax is a rare disorder (1:10 000–15 000 births) with various causes (Noonan's syndrome, chromosomal anomalies, immunological, heart failure, idiopathic), defined by the accumulation of fluid in the pleural cavity. The availability of prenatal diagnosis by ultrasound scan and the emergence of various intrauterine treatment options in fetoscopic thoracocentesis has improved its outcomes. The choice of prenatal therapeutic options depends primarily on the severity of the intrathoracic liquid accumulation and the gestational age of the fetus at diagnosis. Spontaneous resolution of the pleural effusion has been described. However, isolated fetal pleural effusions or hydrothorax generally have a poor outcome, with neonatal death rates varying from 55% when diagnosis is made before 32 weeks gestation to 30% when the diagnosis is made later. Mortality may rise close to 100% when hydrops is associated. Interventions on the fetus aim to drain the intrathoracic fluid by either pleuroamniotic shunt, where one or several shunt devices are left in place, or thoracocentesis, where liquid is evacuated after single or multiple thoracostomy punctures.

If the diagnosis is made in early pregnancy, pleuroamniotic shunting is considered to be the treatment of choice, as thoracocentesis is often followed by rapid restitution of the hydrothorax. The persistence of pleural effusions particularly in the first or second trimester impedes normal development of the lungs, resulting in a significant risk of lung hypoplasia. In contrast, if hydrothorax is diagnosed close to term, the favoured therapeutic approach is by either ultrasound guided fetal thoracocentesis or neonatal transthoracic puncture immediately after birth. However, mortality reported for both fetal intervention and conservative management followed by neonatal intervention remains high at 33% and 37% respectively depending on the cause, associated heart failure, and lung hypoplasia.

Ex utero intrapartum treatment (EXIT) is a management strategy that allows interventions on a neonate delivered from the uterus with the fetoplacental circulation preserved. The aim is to maintain oxygenation in the neonate who has short lived respiratory insufficiency. So far, EXIT has mainly been used for the perinatal management of prenatally diagnosed extrinsic (teratomas, lymphangiomas) or intrinsic (laryngeal atresia, congenital high airway obstruction syndrome) obstructive malformations of the upper airways. Various interventions have been performed with EXIT support including tracheotomy, tracheoplasty, bronchoscopy, intratracheal intubation, surfactant instillation, and, more rarely, ablative surgery of compressive tumours. Most recently, surgical occlusion of the trachea has been advocated for the treatment of fetuses with severe pulmonary hypoplasia secondary to congenital diaphragmatic hernia in order to attempt catch up growth of the hypoplastic lungs and to improve the outcome. In this situation, rapid repermeabilisation of the voluntarily occluded trachea may be undertaken safely during short lived maintenance of the fetoplacental circulation at birth.

We report our experience with EXIT in a fetus presenting in late gestation with severe bilateral pleural effusion.

CASE REPORT

After an initially uneventful pregnancy, this 32 year old woman with type II diabetes was referred to our centre for further assessment of polyhydramnios and suspected macrosomia. Obstetric ultrasound scans up until 32 weeks gestation had been normal.

On the patient's admission at 38 weeks gestation, the fetal scan showed severe polyhydramnios, as well as a moderately macrosomic fetus with isolated bilateral pleural effusions (fig 1). Echocardiographic assessment excluded cardiac malformation, but showed that the umbilical cord vein and inferior vena cava were dilated to internal diameters of 1 cm and 0.65 cm respectively. An additional sign of increased central venous pressure was the reduced peak flow velocity measured by pulsed Doppler within the ductus venosus (0.35 cm/s; normal at term: 0.75 cm/s). No cardiac anomalies were detected, and systolic ventricular function appeared to be preserved. However, the Doppler flow velocity pattern through the atrioventricular valves suggested impaired diastolic ventricular filling (fig 2), probably caused by extrinsic compression of the fetal heart by the extensive fluid accumulation, which occupied the whole intrathoracic space (tamponade effect). On two dimensional ultrasound examination, the lungs appeared to be completely collapsed and echo dense (fig 1). The biophysical profile and cardiotocography were normal.

Three options for management were discussed: thoracocentesis (a) in utero, (b) ex utero intrapartum (EXIT), and (c) postnatally after complete delivery.

Ultrasound guided thoracocentesis before delivery of the fetus is the most commonly used approach in similar fetal pathologies. The right hemithorax is usually drained first to minimise compression of the central veins by the opposite side. In this case, polyhydramnios and the fetal position in cephalic presentation with its back low in the maternal left flank restricted low risk access in utero to the right hemithorax.

Abbreviations: EXIT, ex utero intrapartum treatment.
Delivery of the fetus followed by immediate postnatal drainage of a very large volume of bilaterally accumulated pleural fluid was considered to pose a high risk of prolonged and, possibly severe, hypoxia. Despite immediate intubation and ventilation of the newborn, insufficient lung expansion had to be expected until at least part of the effusion had been withdrawn.

Thus EXIT supported thoracocentesis appeared to be the lowest risk procedure. Incomplete delivery of the fetus by caesarean section was planned. Temporary preservation of the fetoplacental circulation was to allow sufficient time for both thoracic cavities to be drained and the lungs to distend before the newborn had to rely on its own lung function.

A detailed therapeutic plan was set up to minimise time lost during perinatal management. Rachi anaesthesia was chosen for minimal maternal risk, and fentanyl was added during the intervention to provide fetal analgesia. Caesarean section was performed by the obstetric team in the presence of two neonatologists dressed in sterile surgical clothing. Avoiding the inferior margin of the anteriorly placed placenta, uterotomy was performed leaving the uteroplacental and placental fetal circulations intact. A normally developed slightly oedematous baby girl (weight 3670 g (P90); length 52 cm (P90)) was extracted and positioned on a flat sterile surface. Oedematous baby girl (weight 3670 g (P90); length 52 cm)

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Figure 1 Fetal thoracic scan. Transverse view of the fetal thorax and heart at 38 weeks gestation. A massive effusion [*] is evident in both pleural cavities. The left lung lobes appear completely collapsed or compressed (arrows). The right lung is not seen in this view. On the four chamber view, the fetal heart is normal in size and shape.
were withdrawn after 5 and 10 days respectively. Weaned to room air by day 3, the infant was easily extubated directly from high frequency oscillation ventilation on day 5. After uneventful progression of enteric feeding, the girl was finally discharged home at 25 days of age, fully breastfed, with a normal physical examination and a normalised chest radiograph.

**DISCUSSION**

We report our experience in the management of severe bilateral hydrothorax diagnosed in late gestation. A rapidly developing polyhydramnios called for prenatal assessment. Polyhydramnios may occur secondarily because of extrinsic oesophageal compression by extensive bilateral pleural effusions.14

When hydrothorax develops before 27 weeks, normal fetal lung development is often compromised. Furthermore, cardiac and central vein compression may lead to heart failure and intrauterine demise. When diagnosed in mid-trimester, hydrothorax requires a detailed fetal investigation including morphology scans for associated cardiac and extracardiac anomalies, karyotyping, and maternal serological assessment for detection of treatable conditions. Thoracoamniotic shunting with placement of a permanent device should be considered, particularly if there is evidence of compromised lung development and fetal well-being—for example, mediastinal shift in unilateral hydrothorax, signs of cardiac tamponade in bilateral hydrothorax, fetal hydrops.

So far, fetal hydrothorax detected in late gestation has been classically managed by thoracocentesis in utero or immediately after delivery. This represents to our knowledge the first report of perinatal management of severe congenital hydrothorax or chylothorax by EXIT. Maintenance of the fetoplacental circulation ex utero as the sole or main source of oxygenation of a critically affected newborn for a prolonged time requires close and efficient collaboration between anaesthetists, obstetricians, neonatologists, and sometimes surgeons, and the procedure should be carefully planned. Two hardly predictable complications may hamper the success of EXIT. Firstly, intraoperative uterine haemorrhage may need immediate placental delivery; secondly, uterine contraction after delivery of the newborn may restrict uteroplacental perfusion and require halothane administration. Our experience shows that EXIT may be a useful approach especially for the fetus presenting with severe pleural effusions towards the end of gestation and in whom either accurate in utero drainage is technically impossible or unavailable or drainage post partum would result in profound and prolonged hypoxia until sufficient drainage of pleural fluid allowed lung expansion.

Because of the risks of permanent hypoxaemic brain damage if the postnatal drainage fails to establish rapid lung expansion and alveolar gas exchange, intrapartum thoracocentesis is usually preferred.15 However, ultrasound guided intraputine thoracocentesis carries a risk for both the mother and fetus, and lung puncture and withdrawal of large quantities of pleural fluid may induce considerable fetal distress. In addition, the success of the fetal procedure may be limited by the fetal position, as in our case. Thus perinatal evacuation of large pleural effusions during preserved placentofetal circulation may represent a valid and safe treatment option. The technique provides the same advantages as postnatal drainage with the benefit of preserved oxygenation during the intervention. With regard to the intrapartum approach, the EXIT technique facilitates the invasive approach and enables accurate monitoring of the newborn during and after the procedure. Nevertheless, at present, its use should be limited to late gestational interventions in potentially viable neonates.

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