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Percutaneous In Utero Thoracoamniotic Shunt Creation for Fetal Thoracic Abnormalities Leading to Non-Immune Hydrops

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Abstract

Purpose—In a fetus, rare, fetal thoracic abnormalities can cause mediastinal shift and vena cava obstruction resulting in fetal hydrops and intra-uterine fetal demise. This series describes a trans-abdominal, trans-uterine Seldinger based percutaneous approach to create a shunt for treatment of these fetal abnormalities.

Material and Methods—Five fetuses presented with non-immune fetal hydrops due to fetal thoracic abnormalities causing severe mass effect. Under direct ultrasound guidance, an 18 G

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needle was used to access the malformation. Through a peel away sheath, a customized pediatric transplant 4.5 French double J ureteral stent was advanced; the leading loop was placed in the fetal thorax and the trailing end left outside the fetal thorax within the amniotic cavity.

Results—Seven thoracoamniotic shunts were successfully placed in 5 fetuses, with one shunt immediately replaced due to displacement during the procedure and the second not functioning at follow-up requiring insertion of a second shunt. All fetuses had successful decompression of the thoracic malformation, allowing lung re-expansion and resolution of hydrops. Three of 5 mothers had meaningful (> 7 days) prolongation of their pregnancies. All pregnancies were maintained to > 30 weeks, with a range of 30 weeks 1 day to 37 weeks 2 days. There were no maternal complications.

Conclusions—Seldinger based percutaneous approach to draining fetal thoracic abnormalities is feasible and can allow for prolongation of pregnancy, antenatal lung development and ultimately result in fetal survival.

Introduction

Thoracoamniotic shunting has been a well-established treatment for thoracic fetal abnormalities, including fetal pleural effusions, chylothoraces and type I congenital pulmonary airway malformations (CPAMs) since first described in 1988 (1). Mortality is usually due to fetal non-immune hydrops, a severe condition in which excessive fluid accumulates in fetal soft tissue and serous cavities (Figure 1) (2). In the absence of maternal circulating red cell antigens and fetal anemia, this is called non-immune hydrops. There are numerous causes of non-immune hydrops, but it is often seen in cases of intra-thoracic masses, due to mediastinal shift, decreased venous return and severely depressed cardiac output (3). Long term compression of the normal fetal lung tissue can lead to pulmonary hypoplasia and severe neonatal morbidity and death (4). The indication for antenatal intervention is the presence of an isolated large structural abnormality that causes non-immune fetal hydrops in an extremely premature fetus prior to 30-32 weeks gestational age (5-7). The goal of this therapy is to reverse the hydrops, relieve lung compression and prolong the pregnancy, thus avoiding fetal death, reducing the risks of prematurity and allowing for definitive neonatal treatment (8).

Traditionally, thoracoamniotic shunts were created using the trocar technique, which is performed by direct puncture of the fluid collection using a central stylet within a catheter (9). This series describes an ultrasound-guided, trans-uterine Seldinger based percutaneous approach to create a shunt for treatment of fetal thoracic abnormalities as performed by a Maternal Fetal Medicine (MFM)-Interventional Radiology (IR) team.

Materials and Methods

The institutional review board (IRB) approved this retrospective study and waived informed consent. This study was also HIPAA compliant. In addition, a special committee of the IRB was convened in each case in order to grant approval for compassionate treatment. Five consecutive patients underwent creation of a thoracoamniotic shunt between December 12, 2007 and July 7, 2012 (the date of IRB submission).

The fetuses presented with non-immune fetal hydrops due to fetal thoracic abnormalities causing severe mass effect. All of the fetuses underwent detailed ultrasound evaluation to confirm the diagnosis of fetal thoracic abnormality and exclude other anomalies. Amniocentesis was performed to evaluate for chromosomal abnormalities and Toxoplasmosis, Other, Rubella, Cytomegalovirus and Herpes (TORCH) infections. In order to meet inclusion criteria, all fetuses had to have a thoracic abnormality, no infection, omit aneuploidy-redundant and have a normal karyotype. In most cases either the predominant cyst was aspirated and drained by a single puncture or a fetal thoracentesis was performed. The fetuses that had recurrence of their dominant cysts or re-accumulation of their pleural effusions were closely monitored with weekly ultrasounds. All fetuses with type I CPAMs were given beth-methasone in an effort to decrease the size of the CPAM. When fetal hydrops developed, emergent treatment with an in-utero percutaneous thoracoamniotic shunt by the MFM-IR team was offered. The fetus was excluded if an abnormal karyotype, other congenital malformations, or another underlying etiology for the development of non-immune hydrops were found.

Maternal ages ranged from 22-33 years (mean was 27 years), and prior gestations ranged from 0-3, mean 1.4. All fetal thoracic abnormalities were discovered on ultrasound performed at gestational ages ranging from 20 to 24 weeks 2 days. Initial evaluations in the MFM clinic were performed between gestational ages of 23 weeks 4 days and 29 weeks 3 days. At the initial visit, ultrasound confirmed normal amniotic fluid indices (AFIs) in all patients. Three fetuses were found to have type I CPAMs and two had chylothoraces. Type I CPAMs were characterized as macrocystic with a predominant large cyst. Four fetuses were 46XY and one was 46XX. After initial cysts aspiration or thoracentesis, all of the cysts re-accumulated or effusions recurred.

Percutaneous In Utero Thoracoamniotic Shunt Creation

Maternal anesthesia was obtained with an epidural and fetal anesthesia was administered via intra-muscular injection of vecuronium (0.2 mg/kg). Periprocedural antibiotics with cefazolin was administered. Under ultrasound guidance (E-9 Ultrasound, GE Medical Systems, Pewaukee, WI or iU22 Ultrasound, Philips, Andover, MA), an 18 gauge 20 cm Inrad needle (Inrad, Kentwood, MI) was advanced via a percutaneous trans-abdominal, trans-uterine approach avoiding the placenta and umbilical cord into the fetus' thoracic abnormality (Figure 2). A 0.035" Rosen guidewire (Cook Medical, Bloomington, IN) was then coiled within the fetal thoracic abnormality (Figure 3). A 6 French 13 cm peel-away sheath (Cook Medical, Bloomington, IN) was used as a buttress to stabilize the fetal chest wall. Then, a 4.5 French 6 cm double J ureteral stent (Gyrus ACMI, Southborough, MA) was modified by cutting off the trailing loop thereby leaving a single loop at the leading edge for placement in the fetal thorax. This also allowed for a shorter shunt (6 cm) with known formed and unformed lengths. As the modified double J was advanced through the peel-away sheath, it was monitored with ultrasound to ensure that the leading loop was coiled within the fetal thorax and the trailing end was deployed outside the fetal chest wall within the amniotic space. Over the next several minutes, real time ultrasound monitored the collections and confirmed the return of the heart to its midline position. Color Doppler

sonography was used to identify flow through the shunt from the fetal thorax into the amniotic cavity supporting shunt patency (Figure 4).

The mothers were followed with ultrasound and/or MRI weekly to verify shunt function and status of the fetus. The mothers were then discharged home and had weekly follow-up visits in the MFM clinic until delivery. The fetuses were followed up until discharge from the hospital.

Results

A total of 7 shunts were placed in 5 fetuses. The shunts were placed at gestational ages ranging from 25 weeks 1 day to 31 weeks 2 days. There were no maternal complications. Two fetuses required a second shunt to be placed (Table 1). In the first case (# 5), the shunt immediately backed out into the amniotic fluid after the peel away was removed and a second shunt was placed. After the procedure, decompression of the chylothorax was seen; however, on post-operative day 3 the mother went into labor and had premature rupture of membranes, delivering the fetus on post-operative day 4 at 31 weeks 4 days. In the second case (# 4), the first shunt was appropriately positioned initially with decompression of the type I CPAM and the mother was discharged home on post-operative day 2. At her follow-up visit (post-operative day 9), the fluid had re-accumulated and a second shunt was placed on postoperative day 10. Despite a second shunt, the fluid re-accumulated and labor was induced on post-operative day 2 after the second shunt. The fetus was delivered on post-operative day 3 at 31 weeks 5 days.

Four of 5 women maintained normal AFI's after the procedure. In one mother (#1), the ultrasound performed on post-operative day 1 demonstrated profound oligohydramnios. An MRI was performed for further evaluation and the shunt was not seen. There was no fluid within the peritoneal cavity, retroperitoneum, in or surrounding the uterus and no fluid draining from the shunt tract. The mother also denied any leakage of vaginal fluid. A follow-up MRI obtained the following day demonstrated an increase of the AFI to 5.0 cm. Ultrasounds performed on postoperative days 3 and 4 revealed AFI's of 9.0 and 11.7 cm respectively. The shunt was again visualized in the fetal thorax and the effusion was decompressed. Throughout the four day course, no signs of fetal distress were noted. The mother was discharged home on post-operative day 5.

Three of the women were able to go home after shunt creation and mean length of hospital stay was 3 days (range 2-5 days). The remaining two women stayed in the hospital until delivery (#4, due to premature contractions that resulted in delivery 3 days after shunt creation, and #5, due to induction of labor due because of a non-functioning shunt (as mentioned above)). The average dwell time of the fetal shunt was 28 days (range 3-84 days). Three of the 5 mothers had meaningful prolongation of their pregnancies (> 7 days). All fetuses were delivered after 30 weeks gestational age, with ranges of 30 weeks 1 day to 37 weeks 2 days. There was only 1 premature rupture of membranes (PPROM) (#5). One fetus developed fetal distress (#4) and labor was induced. Four of the fetuses were delivered via normal spontaneous vaginal deliveries and one had a Caesarian section due to the mother's

history of prior C-section. All of the shunts were in place in the thoracic cavity after delivery (Figure 5). In #5, the misplaced shunt was found embedded in the placenta.

Three of the infants underwent uncomplicated surgical resection of their type I CPAMs and were discharged home. The two infants with chylothoraces had bilateral chest tubes placed after delivery. Once their effusions resolved, the chest tubes were removed and the infants were discharged home.

Discussion

The quest to treat fetal disease in-utero and avoid delivery with extreme prematurity has been fraught with complications. Even such simple procedures such as amniocentesis have a rare but very real risk of pregnancy loss from rupture of membranes, infection and/or preterm labor and delivery (10). The extreme of fetal treatment with open uterine surgery via hysterotomy, has considerable limitations and obvious maternal and fetal risks (11). Accessing the fetus with minimal breach of the uterus and efforts to minimize maternal risk are paramount to successful treatment and outcome for both patients. Therefore, by forming a multi-disciplinary team, led by MFM and IR, fetal interventions can be approached with minimally invasive techniques in cases where expectant management has a high probability of fetal death.

Thoracoamniotic shunting was first described in the New England Journal of Medicine in 1988 by Rodeck, et al as a method to decompress fetal hydrothoraces allowing more normal development of the fetal lungs, thereby decreasing overall neonatal death (1). More recently, a series of 26 thoracoamniotic shunts in 19 pregnancies were placed in fetuses with pleural effusions or type I CPAMs (9). This series reported postnatal survival rates of 70% for shunts placed in fetuses with pleural effusions and 67% for those with type I CPAMs. Another retrospective study evaluated outcomes in fetuses diagnosed with type I CPAMs in utero and demonstrated an overall survival rate of 75%; however, all the fetuses that developed severe hydrops prior to 30 weeks in their series died (12). In another study, the effectiveness of thoracoamniotic shunting was evaluated in patients with congenital primary hydrothorax and found that neonatal outcomes were improved due to prolongation of pregnancy (13).

In the literature, the most common complication associated with the Trocar technique is displacement of the shunt, which occurred at a rate of 23% (14), compared to this series of 1/7 (14%) displaced. In addition, the reported rate of fetal demise after shunt placement is 38% (13), whereas there were no deaths in our small series. Our series also reports one incidence of oligohydramnios (#1) after shunt creation; however, follow-up MRI and ultrasounds did not demonstrate that the fluid had leaked out into the maternal peritoneal cavity, retroperitoneum, pelvis or subcutaneous tissues. It is possible that the fetus had a mild degree of renal impairment as the result of indomethacin administration or that the fluid was reabsorbed or simply not identified given the large surface area of the maternal peritoneum.

The median duration of gestation after shunt creation by Trocar technique is 2 weeks, with a range of 0-12 weeks (9). In this series, the average dwell time was 4 weeks (range 0-12 weeks). Of the three patients that underwent thoracoamniotic shunting at approximately 25 weeks, all pregnancies were maintained until at least 30 weeks (Table 1).

The trocar technique requires the use of a 3 mm catheter (~9 French) (9, 15). In our Seldinger based technique, an 18 G needle was used to access the fetal thorax. This allows passage of a stiff wire and peel away for a stable system. The modified double J stent accommodates the 0.035" working wire while reducing the diameter by half when compared to the trocar system. Once wire access is achieved, the peel away can be used as a buttress against the fetal thorax and therefore is able to stabilize the fetus prior to placing the stent. Though still invasive, this Seldinger system affords more stability compared to the trocar method, which can be more traumatic to the fetus and allows for very little control. This approach to these lesions is unique, and perhaps affords a decreased likelihood of complications.

Because this is a descriptive study, there are many limitations. It is not known how many women presented to the MFM clinic with non-immune fetal hydrops, but were not candidates for thoracoamniotic shunting because of abnormal karyotypes, concurrent congenital anomalies or harbored fetal infections. Given that these shunts were placed at a large tertiary care referral center, follow-up on the fetuses was limited to their hospitalization.

A Seldinger based percutaneous approach to decompressing fetal thoracic abnormalities is feasible. The new technique affords minimal risk and increases the chance of prolonged in-utero survival with improved outcomes.

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Figure 1. Fetal hydrops

Transverse view of the fetal abdomen obtained as part of a fetal ultrasound. There is a large volume of ascites (black arrow) and skin thickening (calipers) which is consistent with non-immune fetal hydrops.

Figure 2. Thoracoamniotic shunt creation

Fetal ultrasound showing a sagittal oblique view of the fetal thorax. There is a large echogenic left chylous pleural effusion (black arrow). Under direct ultrasound guidance, an 18 G Inrad needle (arrowhead) was percutaneously advanced via a trans-abdominal, trans-uterine approach into the chylothorax. (fetal lung – white arrow, uterus – white arrowhead, amniotic fluid – dashed white arrow)

Figure 3. Thoracoamniotic shunt creation

Transverse view of the fetal thorax demonstrating a guide wire (white arrow) coiled in the fetal pleural effusion (dashed white arrow).

Figure 4. Thoracoamniotic shunt in situ with flow jet

Coronal oblique view of the fetal thorax status post fetal shunt placement. The trailing portion of the shunt is seen within the amniotic cavity (white arrow). Color Doppler images demonstrate a flow jet (white arrow) indicating flow through the shunt into the amniotic fluid (dashed white arrow).

Figure 5. Thoracoamniotic shunt in situ in-utero and postpartum

Transverse view of the fetal thorax with shunt (white arrow) in place and return of the mediastinum to midline. Immediate postpartum photograph demonstrating the in situ thoracoamniotic shunt (black arrow).

Table

Demographic and procedural data for Seldinger based thoracoamniotic shunt creation

Maternal No.	Maternal Age (y)	Fetal dx	Karyotype	GA at presentation to MFM clinic	GA at shunt insertion	GA at delivery	Complications
1	22	B/l Chy	46XX	25w0d	25w2d	37w2d	oligohydramnios
2	23	CPAM	46XY	24w6d	25w1d	30w5d	no
3	33	CPAM	46XY	23w3d	26w3d	30w1d	no
4	26	B/l Chy	46XY	29w3d	29w6d	31w5d	Initial shunt not working, required 2 nd shut
					31w2d		Fetal distress requiring induction
5	31	CPAM	46XY	23w4d	30w6d	31w3d	1 st shunt immediately displaced, 2 nd placed

DX: diagnosis GA: gestational age, MFM: maternal fetal medicine, B/l Chy: bilateral chylothoraces, CPAM: congenital pulmonary airway malformations, (#w/#d): gestational age in weeks/days)