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Thoracoamniotic shunt placement for a right-sided congenital diaphragmatic hernia complicated by hydrops

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ABSTRACT

Mortality associated with congenital diaphragmatic hernia (CDH) is high, and the role of prenatal management continues to evolve. We report a case of a right-sided CDH complicated by fetal hydrops successfully managed with thoracoamniotic shunt placement. Subsequent ultrasounds indicated resolution of hydrops. Despite preterm premature rupture of membrane and preterm delivery at 32 3/7 weeks gestation, the infant survived to hospital discharge at 2.5 months of life. This is the first case of a hydropic right-sided CDH successfully treated with a thoracoamniotic shunt.

Congenital diaphragmatic hernia (CDH) occurs in 1 out of every 2000–4000 pregnancies [1] of which right-sided lesions account for only 10–15% [2]. Few cases of right-sided CDH complicated by hydrops have been reported but when present, the prognosis is dismal [3,4]. It has been suggested that hydrops in patients with right-sided CDH results from caval compression, resulting in reduced preload and impaired cardiac output [4]. When hydrops is seen in association with right-sided CDH, the appropriate management is unclear. Here, we present a hydropic right-sided CDH successfully treated with a thoracoamniotic shunt.

1. Case report

A 32 year old gravida 2 para 1 woman with a previous child diagnosed with hypoplastic left heart syndrome presented with a 24 week ultrasound demonstrating a fetus with a right-sided CDH with bowel, liver, and gallbladder herniating into the right hemithorax. A right pleural effusion, integumentary edema, and ascites were also present, consistent with non-immune hydrops (Fig. 1A). The echogenicity of the liver and large pleural effusion prohibited an accurate lung-to-head ratio calculation. Additional findings included mild placental thickening (3.8 cm), a normal amniotic fluid (amniotic fluid index 17.7), and an estimated fetal weight in the 65th percentile. By report, her 20 week ultrasound had been normal. Fetal echocardiogram revealed no structural cardiac abnormalities but decreased combined cardiac output measured at 98.5 mL/min (<5%ile for estimated gestational age), likely resulting from compression of the vena cava. The following day, a percutaneous fetal thoracentesis was performed, with drainage of 50 mL of straw-colored fluid. Analysis of the pleural fluid revealed a normal female karyotype and negative virology. Fetal cardiac output and integumentary edema improved but the ascites persisted.

At one week follow-up, an ultrasound identified reaccumulation of the right pleural effusion with mediastinal shift, recurrence of the integumentary edema, and less than expected interval growth. Given these findings, a right thoracoamniotic shunt was placed. Under ultrasound guidance, a Harrison catheter was successfully placed through a 3 mm trocar and confirmed to be in adequate position, draining the right pleural effusion (Fig. 1B). There were no immediate fetal or maternal complications.

Serial ultrasounds demonstrated resolution of the right pleural effusion, integumentary edema and ascites as well as improved combined cardiac output (364 mL/min). There was, however, persistent left mediastinal shift from the herniated abdominal contents. At 32 3/7 weeks gestation the patient was admitted

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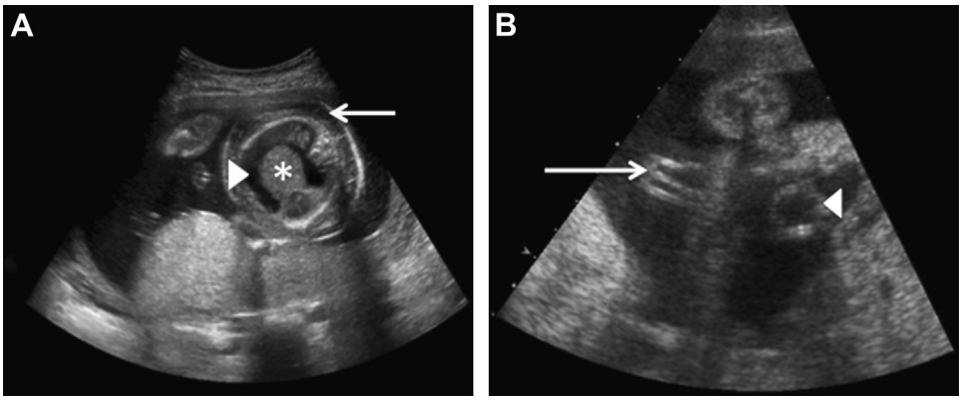

Fig. 1. Fetal ultrasound of right-sided congenital diaphragmatic hernia (CDH). A. Right CDH with liver herniation into the thorax (*) and associated hydrops indicated by a pleural effusion (arrowhead) and diffuse integumentary edema (arrow). B. Ultrasound one day following thoracoamniotic shunt placement, creating a communication between the right pleural cavity (arrowhead) and amniotic cavity (arrow).

for preterm premature rupture of membranes in preterm labor despite latency antibiotics. She had previously received a course of betamethasone. She underwent a spontaneous vaginal delivery of a 2.0 kg female. At delivery, the neonate had an endotracheal and orogastric tube placed in addition to receiving surfactant. On day of life two, primary repair of a right-sided CDH was performed at which point a thin hernia sac was identified. Postoperatively, she required intravenous dopamine, positive pressure ventilation, and inhaled nitric oxide for approximately 2 weeks. Tube feeds were started at 2 weeks of life and reached goal 9 days later. Weekly echocardiograms demonstrated persistent moderate pulmonary hypertension (>50% systemic right ventricular pressure) as well as a patent ductus arteriosus which was closed percutaneously at 2 months of age. She was discharged home at age 2.5 months on 1 L oxygen by nasal cannula and bosentan for continued treatment of persistent pulmonary hypertension.

2. Discussion

The mortality associated with right-sided CDH is significant and is reported to be between 30 and 45% [5,6]. The “hidden mortality” [7] was described to account for the high prenatal mortality rate with at least 10% suffering in utero fetal demise. The role of prenatal management for fetuses with a CDH is an area of intense investigation. Currently, prospective trials are ongoing to evaluate the efficacy of fetal balloon tracheal occlusion, which appears to be a promising prenatal therapy to improve outcomes for CDH. However, this application is intended to treat pulmonary hypoplasia and hypertension, not hydrops.

Hydrops is a rare complication of CDH but is associated with near uniform mortality in patients with CDH [3]. Therefore, when hydrops develops, fetal intervention may be life-saving. For the fetus with a right-sided CDH who develops hydrops close to term, delivery may be best practice, although percutaneous thoracentesis may be a therapeutic alternative. Management is more controversial in fetuses presenting earlier in gestation. This is the scenario in which placement of a thoracoamniotic shunt has the potential to be life-saving.

In our case, a thoracentesis was initially performed with re-accumulation of the pleural effusion within 1 week. Our preference has been to perform a thoracentesis prior to proceeding to thoracoamniotic shunt placement. We and others have observed that a single thoracentesis can be definitive without re-accumulation of the pleural effusion which avoids the need for a more invasive procedure such as a thoracoamniotic shunt [8]. Thoracentesis also provides a window to assess how the fetus responds physiologically to decompression of the effusion prior to subjecting the mother and fetus to a more invasive procedure. Additionally, chromosomal analysis can be obtained from pleural fluid aspirate which can facilitate counseling, given the poor outcomes when abnormalities are associated with CDH [9]. Multiple complications have been reported with thoracoamniotic shunt placement including fetal hemorrhage from intercostal artery laceration, shunt dislodgement or plugging, chorioamnionitis, membrane separation, preterm delivery, and fetal demise [10]. Moreover, this patient’s preterm delivery was likely related to the combination of fetal hydrops and fetal intervention.

We hypothesize that these cases are affected by an initial insult of caval compression from herniated viscera that leads to the slow progression of heart failure, coupled with progressive accumulation of transudative fluid in the right chest. Eventually, this large pleural effusion causes further caval compression and exacerbates the mild cardiac failure, resulting in hydrops. Interestingly, this patient had a small diaphragmatic defect that was repaired primarily, whereas hydrops in CDH is often associated with larger defects [3]. We have recently reported the close correlation between prenatal inflammatory mediators and postnatal pulmonary hypertension in patients with CDH [11]. This patient’s unusual combination of a small diaphragmatic defect with hydrops and persistent pulmonary hypertension suggest a more severe disease process than would be expected by the size of the defect. Further investigation into the mechanisms of the development of hydrops in CDH is necessary.

3. Conclusion

While development of hydrops in right-sided CDH portends a grim prognosis, our experience suggests that thoracoamniotic shunt placement for right-sided CDH complicated hydrops is not only feasible but can be life-saving.

Conflict of interest statement

The authors have no conflict of interest to report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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