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Repair of a bilateral Morgagni hernia in a premature, extremely low birth weight infant

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ABSTRACT

Congenital diaphragmatic hernias (CDH) are rare congenital anomalies occurring in 1:5000 live births with a wide range of clinical presentations. The Morgagni hernia is the rarest of these defects with the hernia presenting anteriorly, most commonly on the right. Factors found to predict mortality in infants presenting with CDH include prenatal diagnosis, early gestational age, low birth weight, large diaphragmatic defect, associated congenital anomalies, and low APGAR scores. We present the case of a 27-week gestational age premature girl weighing 460 g who required urgent repair of a Morgagni diaphragmatic hernia and a review of the current literature on the topic. To the best of our knowledge, this is the smallest infant to undergo repair of this CDH reported to date.

Congenital diaphragmatic hernias (CDH) are rare, occurring in 1:2000 to 1:5000 live births [1]. The rarest CDH is the Morgagni hernia, with an overall incidence of 3%–4% among diaphragmatic hernias [2]. A Morgagni hernia is thought to occur due to lack of fusion or muscularization of the pars sternalis and pars costalis anteriorly during diaphragmatic development leading to a triangular parasternal gap known as the Morgagni gap on the right and the Larrey gap on the left [2,3]. About 90% of Morgagni hernias occur on the right, 2% on the left, and 8% bilaterally [2]; a hernia sac is present in more than 95% of cases [1].

The clinical presentation of these defects is variable and can range from severe respiratory distress in the neonate to an incidental finding in adulthood. Common presentations later in life include recurrent respiratory infections and gastrointestinal complaints [1]. While Morgagni hernias have been found in association with congenital abnormalities including Down syndrome, Pentalogy of Cantrell, Noonan syndrome, Prader–Willi syndrome, Turner's syndrome, and intestinal malrotation [1,2], there is no evidence of a genetic cause for CDH. Most infants diagnosed with CDH are full-term and about half are diagnosed prenatally. Many factors have been found to predict mortality in infants with CDH including prenatal diagnosis, low birth weight, early gestational age, large diaphragmatic defect, and low APGAR scores. Prematurity is considered a significant contributor to mortality and is associated with close to 50% mortality, an effect that is lessened with increasing gestational age [4,5].

We present the case of a 27-week gestational age (GA) 460 g female infant requiring urgent repair of a bilateral Morgagni hernia in the setting of worsening respiratory status on DOL 14 and a review of the current literature. Despite the patient's prematurity and size, she demonstrated an immediate physiologic improvement with the repair and was ultimately discharged home without the need for supplemental oxygen. With two year follow-up, there is no evidence of recurrence and she continues to grow and develop normally. To date, this is the smallest infant with repair of a Morgagni congenital diaphragmatic hernia reported in the literature.

1. Case report

A 27-week, 460 g female was born via emergency cesarean section for non-reassuring fetal heart rate to a 35 year old G1P0 mother. The pregnancy was complicated by pre-eclampsia with

Abbreviations: CDH, congenital diaphragmatic hernia; GA, gestational age; DOL, day of life; HELLP, hemolysis elevated liver enzymes low platelets; HFOV, high frequency oscillatory ventilation; NICU, neonatal intensive care unit.

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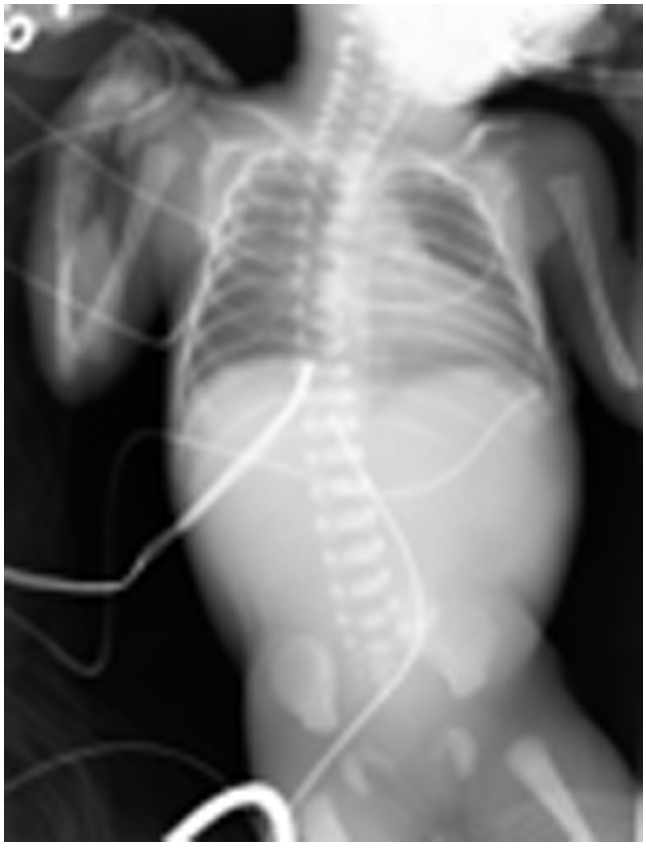

Fig. 1. Initial chest X-ray. The X-ray demonstrates cardiomegaly, mild interstitial prominence of lungs, and a gasless abdomen.

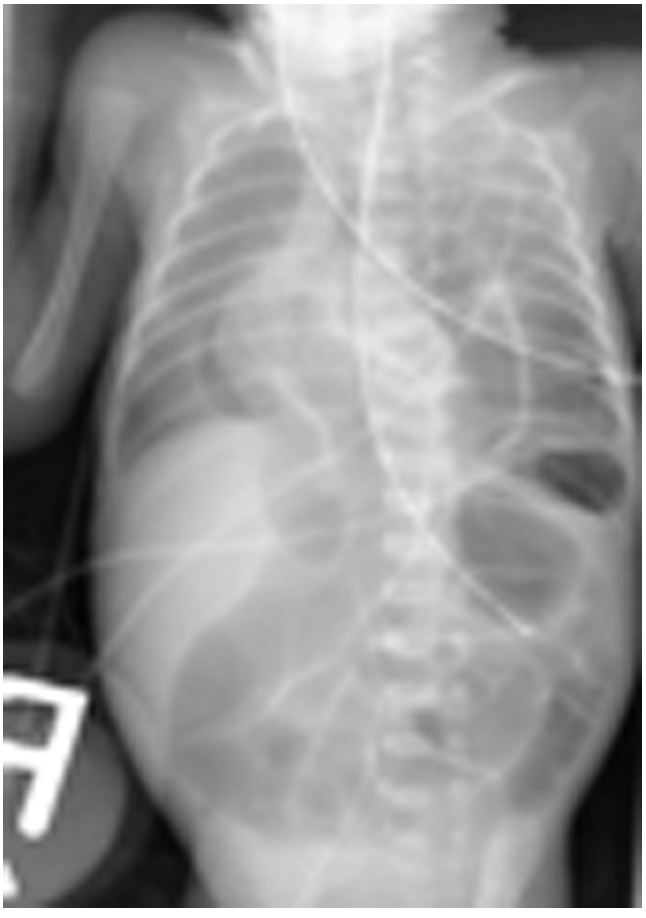

Fig. 2. Abdominal X-ray (DOL 5). Abdominal X-ray demonstrating herniation of abdominal contents into the left hemithorax and overlying the heart.

hemolysis, elevated liver enzymes, low platelets (HELLP) syndrome and intrauterine growth restriction. The prenatal ultrasound was unremarkable. Her APGAR scores were 4 at 1 min and 8 at 5 min. She required intubation with positive pressure ventilation and was admitted to the NICU. The patient was placed on high frequency oscillatory ventilation (HFOV) and an initial chest X-ray is shown (Fig. 1).

The patient was extubated to continuous positive airway pressure (CPAP) on DOL 3. After enteral feeds were initiated, abdominal distention was noted and an abdominal radiograph demonstrated distended loops of bowel without pneumatosis or free air. On DOL 5, the abdomen remained distended and discolored. A repeat X-ray revealed herniation of abdominal contents into the left hemithorax and overlying the right heart (Fig. 2). The patient was re-intubated with a gradual increase in the ventilatory support required over the ensuing days.

On DOL 14, the patient's condition worsened rapidly and she required maximal ventilatory support. A chest radiograph obtained at this time demonstrated an increased amount of distended bowel filling the right and left thorax (Fig. 3). Despite 100% inspired oxygen, paralysis, and HFOV, she was unable to maintain her oxygen saturation with a capillary blood gas of 7.32/43/43.3/21/86%/-3.6. Since her rapid clinical deterioration appeared to correlate with the dramatic increase in the intrathoracic intestinal distention, she underwent an emergency repair of the diaphragmatic defect in the neonatal intensive care unit.

The patient was 625 g at the time of operation. Through a transverse laparotomy, a large amount of normally rotated, viable bowel was found in the thorax with a hernia sac present. The entire bowel was evaluated and found to be of normal length and caliber. A primary closure of the anterior bilateral diaphragmatic defect, measuring approximately 2 cm at its greatest diameter, was performed (Fig. 4). Due to the tenuous respiratory status and the significant abdominal pressure required to primarily close the abdominal cavity, a 3 cm silastic spring-loaded silo (Bentec Medical, Woodland, CA) was utilized as a temporary closure (Fig. 5). Immediately, the patient's respiratory status dramatically improved and required significantly less ventilator support. She was placed on Synchronized Intermitted Mandatory Ventilation Pressure Limited Time Cycled Continuous Flow with a PIP 17, PEEP 6, Rate 40, and FiO_2 of 25% and a capillary blood gas of 7.27/46/52/19/88%/-5.7. On DOL 21 (POD 5), fascial closure with a 2 cm by 4 cm AlloDerm (LifeCell, Bridgewater, NJ) regenerative tissue patch was accomplished with a skin defect permitted to close by secondary intention (Fig. 6).

The postoperative hospital course was complicated by acute pulmonary hemorrhage treated with FFP, elevation in PEEP, and epinephrine given via endotracheal tube on DOL 33 and bacteremia treated with antibiotics on DOL 42. Additionally, she was treated with steroid therapy for chronic lung disease. There was no evidence of pulmonary hypertension on an echocardiogram performed on DOL 55 with the patient successfully weaned to room air by DOL 66. She was discharged home weighing 2296 g, on room air, tolerating oral feedings, and with a well-healed incision site on DOL 128.

At 1-year follow-up, the patient continued to grow and develop with a height and weight of 61.6 cm and 5.6 kg (less than the 5th percentile for height and weight for her gestationally corrected