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 [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, 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.

1. Case report

A 27-week, 460 g female infant 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...
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 Intermittent Mandatory Ventilation Pressure Limited Time Cycled Continuous Flow with a PIP 17, PEEP 6, Rate 40, and FiO2 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...
At 2 year follow-up, she does not require oxygen and has had no additional hospitalizations. There is no symptomatic or radiographic evidence of recurrence of the Morgagni hernia (Fig. 7).

2. Discussion

Congenital diaphragmatic hernias are rare and challenging malformations. Studies have focused on the more common Bochdalek, posterior diaphragmatic hernia, with little information focused on premature infants with the rare Morgagni hernia. The Morgagni hernia has an overall incidence of 3%–4% among diaphragmatic hernias. Ninety percent are found on the right side, 8% are bilateral, and 2% are on the left [2]. These retrosternal diaphragmatic defects have vague and variable presentations. The majority of patients are asymptomatic and are rarely diagnosed in the neonatal period [6]. When diagnosed in infants and children, presenting symptoms include respiratory distress, recurrent pneumonia, vague gastrointestinal symptoms, or general epigastric discomfort with vomiting or coughing [1,6]. Commonly herniated organs include the colon, small bowel, liver, omentum, and stomach [1,7]. Acute cases can present with intestinal ischemia secondary to strangulation within the defect or gastric volvulus [6]. The defect may also be identified incidentally during the evaluation for an associated congenital anomaly [1]. A Morgagni hernia is usually diagnosed with a chest X-ray demonstrating air-fluid levels in the midline of the chest and a hernia in the retrosternal space on lateral views. Small hernias may require contrast radiography or cross-axial imaging to confirm the diagnosis [1,6].
shifted the management of CDH away from emergent repair and toward early stabilization, improving ventilator strategies, and reducing barotrauma through gentle ventilation and permissive hypercapnia [12,13]. Extracorporeal membrane oxygenation (ECMO) is also often employed in the management of CDH although its efficacy is still unclear [13,14].

Unlike Bochdalek hernias, Morgagni hernias usually are not associated with substantial lung hypoplasia. However, they may be associated with pericardial, sternal, and abdominal wall defects as part of the Pentalogy of Cantrell [15]. Most authors recommend surgical repair of a Morgagni hernia, even in patients who are asymptomatic, relatively soon after diagnosis due to the risk of incarceration and strangulation [1,6]. A laparoscopic or open transabdominal approach is favored over a transthoracic approach due to the high incidence of bilateral hernias and associated gastrointestinal anomalies that can be evaluated using the transabdominal approach [6,7].

The patient discussed in this report is very unusual in that she presented very early in life with a symptomatic Morgagni hernia. She was in acute respiratory distress and her clinical status deteriorated as the hernia contents occupied additional space in her thorax. She appeared to benefit immediately from emergent decompression of the thoracic contents suggesting that the intestinal distension in the thorax directly compromised cardiopulmonary function. Emergent surgical repair is not the usual treatment for patients with respiratory distress secondary to a Bochdaleck CDH since the underlying pathophysiology is related to lung hypoplasia and pulmonary hypertension rather than a space-occupying lesion. ECMO was not technically feasible for the 625 g infant presented in this case study, leaving surgical exploration as the last resort. Although our patient had many risk factors associated with a poor outcome in CDH, it is noteworthy that the diaphragmatic hernia was not identifiable on radiographic studies prior to DOL 5. This suggests that perhaps the CDH did not impede pulmonary development in utero, thereby minimizing the degree of pulmonary hypoplasia and pulmonary hypertension. Repair of this lesion in a neonate with worsening respiratory status not attributable to pulmonary hypertension led to rapid and dramatic improvement. This is very different from the common underlying pathophysiology of the more common Bochdaleck hernia in which the repair usually does not lead to immediate physiologic improvement.

3. Conclusion

To date, this is the smallest infant to survive a Morgagni CDH repair reported in the literature. Despite an early gestational age, an extremely low birth weight, and a rare bilateral Morgagni hernia, the patient survived to hospital discharge and is free of recurrence with 2 years of follow-up. Despite reported poor outcomes with premature infants with CDH, the prognosis for patients with a Morgagni CDH may not correlate with our more common understanding and expectations for newborns with a Bochdaleck CDH.

References


Fig. 6. Intraoperative image abdominal closure (DOL 21). Fascial closure with a regenerative tissue patch.

Fig. 7. Chest X-ray at 1 year follow up. Chest X-ray taken at 1 year follow up.


