

Hepatopulmonary fusion associated with right-sided congenital diaphragmatic hernia: management of this rare anomaly and a review of the literature

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Abstract

Hepatopulmonary fusion is a rare malformation that is often discovered during operative repair of right-sided congenital diaphragmatic defects. Based on a search using medical search engines, we only found 22 cases of hepatopulmonary fusion in the English literature worldwide to date. We describe herein a case of hepatopulmonary fusion with right-sided congenital diaphragmatic hernia in a female neonate who presented with respiratory distress. We discuss management of this case and review the relevant literature.

Keywords

Congenital diaphragmatic hernia, hepatopulmonary fusion, respiratory distress, neonate, liver, lungs, diaphragm

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Introduction

Right-sided congenital diaphragmatic hernia (CDH) accounts for approximately 15% of CDH cases. Patients with right-sided CDH typically do not have symptoms at birth. Therefore, presentation and diagnosis of CDH are often delayed for such patients.^{1–3} The liver is the most commonly

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herniated organ for right-sided defects. On rare occasions, the herniated liver fuses with the hypoplastic right lung, causing hepatopulmonary fusion (HPF), which is often discovered during operative repair of the defect. We describe here a case of HPF with right-sided CDH and its management, and review the relevant literature.

Case Report

A full-term female neonate was admitted to neonatal intensive care immediately after birth with respiratory distress and referred to surgical service on the 2nd day. At that time, she was haemodynamically stable and maintained on 2 L of oxygen. Her breathing sounds were decreased on the right side. A chest radiograph showed opacification of the right hemi-thorax with a mediastinal shift (Figure 1). Ultrasound confirmed the presence of right-sided CDH and showed that the liver had herniated through the defect (Figure 2). An ultrasound of the abdomen and echocardiography were normal. These examinations were performed to check for other associated anomalies, and to assess cardiac function and pulmonary pressure. After 48 h of life, operative repair was planned for the patient. On exploration, a right posterolateral diaphragmatic defect (Bochdalek hernia) was found, containing the right lobe of the liver, the right kidney with its suprarenal gland, and a loop of the colon. The liver, particularly its posterior aspect, was densely adherent to the diaphragm and chest wall. The liver was abnormally lobulated and firmly fused with the hypoplastic ipsilateral lung. Throughout the dissection, no distinct plane between the lung and liver could be established, even after prolonged effort. During the dissection, a tear in the inferior vena cava occurred near its junction with the hepatic vein. This tear was immediately controlled and repaired. The liver, colon, and kidney with its suprarenal



Figure 1. Chest X-ray shows left side opacity with mediastinal shift to the right side



Figure 2. Ultrasound image showing a herniated liver inside the chest

gland were placed back in the abdomen, and repair of the diaphragmatic defect was performed. A portion of the liver that we failed to separate from the lung remained, and the diaphragmatic defect was repaired around it without causing any tissue compression. The patient was moved to the neonatal intensive care unit and intubated and ventilated. Unfortunately, during the postoperative period, the patient persistently exhibited respiratory failure, pulmonary hypertension, and hemodynamic instability. She died on the third postoperative day.

Verbal consent for publication was obtained from the parents.

Discussion

HPF is a rare malformation in patients with right-sided CDH. The spectrum of HPF ranges from only fibrovascular communication to complete parenchymal fusion that can be difficult to divide.⁴ We describe one case of HPF with right-sided CDH that involved tissue fusion between the lower lobe of the lungs and the superior surface of the liver. A review of English literature using the search engines PubMed, Scopus, and Google scholar database was performed by searching with the following keywords: congenital diaphragmatic hernia, hepatopulmonary fusion, hepatic pulmonary fusion, and liver and lung fusion. We also searched references in the retrieved articles to identify other reported cases. Only 22 cases of HPF were identified (Table 1).⁴⁻¹⁸ The embryological basis for development of HPF is not yet understood. One theory for this development is that failure in formation of the diaphragm during early gestation allows hepatic tissue to herniate into the chest during the 10th week of gestation. Subsequently, the liver may fuse to the primitive lung. Another theory is that HPF is the primary anomaly that then

interferes with complete development of the diaphragm.⁵

The clinical presentation of HPF is that of Congenital diaphragmatic hernia, which is commonly discovered during the early neonatal period, when respiratory distress is the dominant feature. However, asymptomatic cases of HPF at birth present late during infancy and are discovered incidentally in a chest X-ray performed for other reasons.⁶ However, HPF is typically not diagnosed preoperatively and is most often discovered during surgical exploration.⁷ HPF should be suspected preoperatively when there is opacification of the right hemi-thorax without a mediastinal shift to the contralateral side. This lack of shifting of the mediastinum is believed to be related to the presence of a hypoplastic lung on the affected side.⁴ Exceptions to the above-mentioned description of HPF can occur. Mediastinal shift may be detected in HPF cases if the defect is large and the bowel and other organs are herniated into the chest, as observed in our case and described in other reports.^{4,6} With routine use of ultrasound in antenatal care, CDH can be diagnosed antenatally. Ultrasonographic features suggesting right CDH include the absence of bowel loops in the abdominal cavity, bowel loops in the chest, and presence of the liver inside the chest. Moreover, when a colour Doppler study is added, it may show the umbilical segment of the portal vein bowing towards the left and portal branches to the lateral segment of the left hepatic lobe in the direction of or above the diaphragm. Additionally, the gallbladder is observed above the diaphragm, and an echogenic space representing the left hepatic lobe is found between the left border of the heart and the stomach.¹⁹⁻²¹ Although CDH can be diagnosed by ultrasound, the diagnosis of HPF is difficult prenatally. However, with increased use of foetal magnetic resonance imaging (MRI) as a diagnostic tool

Table 1. Previously reported cases of hepatopulmonary fusion

Study	Patients (n)	Sex/age	Associated findings	Outcome at report time
Katz et al. (1998) ⁹	1	Sex not mentioned/ full-term neonate	Right congenital diaphragmatic hernia	Died
Slovis et al. (2000) ⁴	6	Two males and four females/neonates	Five patients had systemic arterial and venous circulation to a fused lung, two had left-sided congenital heart disease, and two had sequestrations	Two patients died Four patients survived
Keller et al. (2003) ⁵	1	Male/full term neonate	Right diaphragmatic hernia	Survived
Robertson et al. (2006) ⁸	1	Male/neonate	Right diaphragmatic hernia	Died
Taide et al. (2010) ¹⁰	1	Sex not mentioned/ 7 months	Right diaphragmatic hernia	Survived
Gander et al. (2010) ⁶	1	Male/3 months	Right diaphragmatic hernia	Survived
Chandrashekhara et al. (2011) ¹¹	1	Male/11 years	Pulmonary sequestration receiving arterial supply from the descending aorta and celiac trunk	Survived
Castle et al. 2011 ¹²	1	Male/neonate	Right-sided congenital diaphragmatic hernia, duodenal atresia, and an imperforate anus	Survived
Hamilton et al. (2014) ¹³	1	Male/3 months	Anomalous right pulmonary venous return, and azygos continuation of the inferior vena cava	Died
Lin et al. (2012) ¹⁴	3	Male/8 months Female/6 years Female/neonate	Two patients with diaphragmatic hernia and one with pentalogy of Cantrell	Survived Survived Died before the operation
Saurabh et al. (2013) ¹⁵	1	Male/neonate	Right-sided diaphragmatic hernia, thumb and index finger syndactyly, and multiple clefts in the vertebrae	Died
Olenik et al. (2014) ⁷	1	Male/neonate	Right diaphragmatic hernia	Survived
Laamiri, et al. (2016) ¹⁶	1	Male/neonate	Right diaphragmatic hernia	Died
Jain et al. (2017) ¹⁷	1	Female/2 months	Right diaphragmatic hernia	Died
Takezo et al. (2017) ¹⁸	1	Female/ diagnosed at 33 weeks' gestation. Operated on at 1 day old	Right diaphragmatic hernia	Not mentioned

for CDH once suspected on prenatal ultrasound, HPF can be identified before delivery and thus before postnatal surgical exploration. Postnatally, MRI is still the diagnostic modality of choice for HPF. Demonstration of venous drainage from the lung parenchyma into the intrahepatic part of the inferior vena cava could be a diagnostic indicator of HPF.⁵ Echocardiography is usually performed to assess cardiac function, associated anomalies, and features of pulmonary hypertension, but demonstration of hepatic veins draining directly into the right atrium also indicates diagnosis of HPF. In our case, neither computed tomography nor MRI was performed because we did not consider HPF, and only ultrasound was performed.

There is no consensus about the optimal time for repairing CDH and no strong evidence to favour an early (within the first 24 h) or delayed approach (after period of stabilization).²² However, an early operation in the presence of compromised respiratory and unstable haemodynamic parameters is unsafe, and delayed repair may allow stabilization of borderline patients and thus improve prognosis.²³ Nevertheless, there are no agreed criteria to define physiological stabilization. Some authors have defined these criteria as stable mean blood pressure for gestational age, preductal oxygen saturation in the range of 85%–95% on $\leq 50\%$ oxygen, urine output >2 cc/kg/h, and serum lactate levels <3 mmol/l.²⁴ Pulmonary artery pressure also determines stability. Some surgeons operate only if normal measures are maintained for at least 24 to 48 h based on echocardiography because they believe that pulmonary hypertension is a major risk of mortality. However, Rozmiarek and colleagues related mortality of CDH to associated complications (cardiac defects and renal failure) and initial blood gases rather than the timing of repair.²⁵ In our case, we preferred to perform immediate repair because the neonate

was haemodynamically stable and pulmonary pressure did not cause a severe shunt.

The aim of surgical repair in HPF cases is to reduce the herniated abdominal content and suture the diaphragmatic defect, which are difficult in cases involving inseparable lung and hepatic tissues. Many procedures have been described, including separation of the lungs and liver with repair of the defect, repair of the defect around the fused organs without separation, and partial hepatectomy and/or pneumonectomy.^{4,6,26} To achieve successful separation of the lung and liver, Robertson et al.⁸ used a Liga-Sure device because blunt dissection causes major air leakage.^{4,8} The prognosis of patients with HPF is poor, and thus patients often die during the perioperative period. High mortality among patients with HP is related to complications of lung hypoplasia, CDH and/or surgical procedures for HPF. These complications include respiratory failure, persistent pulmonary hypertension, right heart failure, congenital heart diseases, and thrombosis of the inferior vena cava.^{6,7} Table 1 shows that 12 of the reported patients survived, while nine children died.

We recommend that the possibility of HPF should be considered in any case of right-sided diaphragmatic hernia. Surgical management of such cases should not be rushed. When feasible, these cases should be assessed via a thorough investigation that invariably includes ultrasonography, a contrast computed tomography scan and/or MRI. This is because the majority of reported cases were discovered intraoperatively.¹⁷ Additionally, the only surrogate indicator mentioned in these previous reports was the absence of mediastinal shift, which was believed to be related to pulmonary hypoplasia. Furthermore, some cases of HPF were associated with mediastinal shift when the defect was large and there were intrathoracic herniated bowels.

Before surgery for HPF in right-sided CDH, careful preparation and optimal stabilization are required. Additionally, all logistical preparations for a major surgical undertaking, potentially including hepatic or pulmonary lobectomy or pneumonectomy, should be performed.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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