Journal of Pediatric Surgery 55 (2020) 1903–1907

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Hepatopulmonary fusion: A rare variant of congenital diaphragmatic hernia☆

Dalya M. Ferguson [a,b,*]
for the Congenital Diaphragmatic Hernia Study Group

[a] Department of Pediatric Surgery and Center for Surgical Trials and Evidence-based Practice (C-STEP), University of Texas John P. and Katherine G. McGovern Medical School 6431 Fannin Street, MSB 5.256, Houston, TX 77030, United States
[b] Children's Memorial Hermann Hospital, 6411 Fannin Street, Houston, TX 77030, United States

ARTICLE INFO

Article history:
Received 20 July 2019
Received in revised form 15 September 2019
Accepted 16 September 2019

Key words:
Case series
Congenital diaphragmatic hernia
Congenital pulmonary venolobar syndrome
Hepatopulmonary fusion
Neonatal

ABSTRACT

Background: Hepatopulmonary fusion (HPF), a rare anomaly associated with right congenital diaphragmatic hernia (CDH), is characterized by a fibrovascular fusion between herniated liver and lung parenchyma. We aimed to clarify patient characteristics, management strategies, and outcomes in HPF.
Methods: Data on infants with HPF were obtained from the Congenital Diaphragmatic Hernia Registry (CDHR). Patient characteristics, management, and outcomes were compared with the results of a literature review.
Results: Ten cases of HPF were identified in the CDHR. Five patients survived. The median estimated gestational age was 38 weeks (range 36–40). Median birth weight was 2.7 kg (range 2.0–3.8 kg), but non-survivors had a lower median birth weight (2.3 kg vs. 3.5 kg). All patients had at least 1 congenital anomaly in addition to CDH. Operative approach varied, but most surgeons performed only partial separation of the liver and lung (n = 6). The 2 patients who underwent complete separation both ultimately died, 1 due to significant postoperative complications and 1 due to severe pulmonary hypertension with multiple vascular anomalies.
Conclusion: Partial separation of liver and lung appears to be the wisest surgical approach in HPF, as complete separation has resulted in catastrophic complications due to frequent underlying vascular anomalies.
Level of evidence: IV.

Hepatopulmonary fusion (HPF), a rare anomaly that may occur in patients with right-sided congenital diaphragmatic hernia (CDH), is characterized by fusion between herniated liver and lung parenchyma. The degree of fusion in HPF has been described to range from fibromuscular communication to complete parenchymal fusion [1]. The ideal management strategy for HPF has not been well-defined due to the rarity of the condition.

Since its establishment in 1995, the CDH Study Group (CDHSG) have collected data on over 12,000 infants from 83 centers around the world in the CDH Registry (CDHR) [2]. The CDHR provided a unique opportunity to describe this rare condition within a large cohort of CDH patients. This series of 10 patients represents the largest HPF case series to date.

1. Materials and methods

We performed a retrospective review of a prospective cohort. In 2015, the CDHR was updated to include a question on the presence or absence of HPF. Several patients born prior to 2015 were also known to have HPF, due to comments included in registry submissions. Data on all patients with HPF were obtained from the CDHR. Additional data, including operative descriptions and imaging results, were requested from the reporting institutions. All contributing authors and institutions are listed in Appendix A. The University of Texas Health Science Center at Houston Institutional Review Board approved this study (HSC-MS-03-223).

A systematic literature search was performed using PubMed. All case reports, case series, and literature reviews available in English were included. References of each publication were reviewed in order to identify additional articles. Data on patient demographics, preoperative status, operative details, and outcomes were collected when available.

Data from the case series and the literature review were 1summarized using frequency distributions for categorical variables and medians with full or interquartile ranges for continuous variables. The number of patients with available data were reported for each

Abbreviations: CDH, Congenital diaphragmatic hernia; CDHR, Congenital Diaphragmatic Hernia Registry; CDHSG, Congenital Diaphragmatic Hernia Study Group; CT, Computed tomography; DOL, Days of life; ECMO, Extracorporeal membrane oxygenation; EGA, Estimated gestational age; HPF, Hepatopulmonary fusion; IVC, Inferior vena cava; LOS, Length of stay; MRI, Magnetic resonance imaging.

☆ Declarations of interest: None.

* Corresponding author.
E-mail address: dalya.m.ferguson@uth.tmc.edu.

https://doi.org/10.1016/j.jpedsurg.2019.09.037
0022-3468/

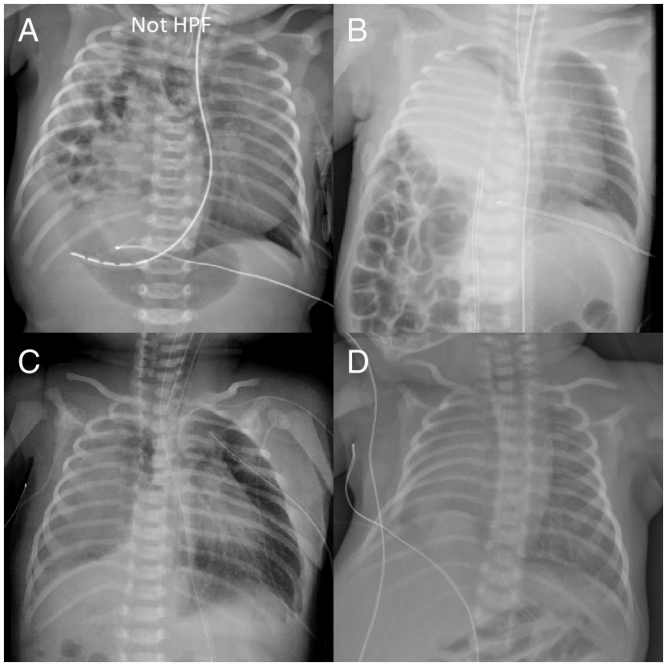

Fig. 1. Chest radiographs. A) Neonate with right-sided congenital diaphragmatic hernia without hepatopulmonary fusion (HPF): bowel and liver in the right hemithorax with significant leftward mediastinal shift. B) Female neonate with HPF: liver and bowel in the right hemithorax with slight leftward mediastinal shift. C) Male neonate with HPF: opacification of the right hemithorax without mediastinal shift. D) Male neonate with HPF: opacification of right hemithorax with rightward mediastinal shift.

variable. Stata version 15 was used for all analyses (StataCorp, College Station, TX).

2. Results

Seventeen potential cases of HPF were identified in the CDHR. Of those, 5 patients were determined not to have HPF and 2 were excluded due to lack of evidence (operative or imaging reports) needed to confirm the diagnosis. This yielded a cohort of 10 HPF patients (Online Supplement 1). Of these, 1 patient was diagnosed with HPF via computed tomography (CT) and has never required surgery (Fig. 1D). However, we included the patient in the cohort because advanced imaging has correctly diagnosed HPF in the past (see Discussion).

Five of 10 patients survived. The majority of patients were born full term with a median estimated gestational age (EGA) of 38 weeks (range 36–40, Table 1). The median birth weight was 2.7 kg (range 2.0–3.8 kg), but non-survivors had a lower median birth weight (2.3 kg vs. 3.5 kg). All patients had at least 1 congenital anomaly in addition to the CDH. Diaphragm defect size, defined by CDHSG grade, was not observed to predict survival.

Most patients were intubated within 24 h after birth (n = 7, Table 2). Extracorporeal membrane oxygenation (ECMO) was utilized in 3 patients, only 1 of whom survived. The 2 patients who underwent CDH repair while on ECMO support did not survive.

Among the 9 patients who underwent repair, the approach to separating the liver and lung varied. Complete separation was achieved in 2 patients. One of them, a male term infant, experienced postoperative bleeding requiring immediate salvage laparotomy with packing. He subsequently developed anuria and expired on postoperative day 3 (day of life [DOL] 9) due to renal failure. A female infant born at 36 weeks EGA also underwent complete separation. Her operation and immediate postoperative course were uneventful, however she suffered from persistent, severe pulmonary vein stenosis and progressively worsening pulmonary hypertension. The patient was evaluated for surgical correction or transplant, but was determined not to be a candidate for either procedure. She was ultimately discharged home with palliative care and expired on DOL 335.

Separation was not attempted in 1 female infant (Fig. 1B). Instead, the pleura and peritoneum that were present around the area of fusion were plicated in order to maximize intrathoracic space. The patient survived to hospital discharge but remained ventilator dependent, and she ultimately expired on DOL 255.

In 6 cases, only partial separation of liver and lung was achieved. Separation was attempted in 1 male patient, but was found not to be feasible due to parenchymal bleeding, as well as a large vessel

Table 1
Patient demographics.

	Non-survivors	Survivors
	n = 5	n = 5
Female, n (%)	3 (60)	2 (40)
EGA in weeks, median (range)	37 (36–40)	39 (37–40)
Birth weight in kg, median (range)	2.3 (2.0–2.7)	3.5 (2.7–3.8)
Cardiac anomalies, n (%)	4 (80)	5 (100)
Major	1 (20)	3 (60)
Vascular anomalies, n (%)	3 (60)	4 (80)
Right lung vascular anomalies	3 (60)	3 (60)
Right pulmonary artery hypoplasia	3 (60)	1 (20)
Chromosomal anomalies, n (%)	1 (20)	0
Other anomalies, n (%)	2 (40)	4 (80)
CDH side, n (%)		
Right	4 (80)	5 (50)
Bilateral	1 (20)	0
Defect type, n (%)		
B	2 (40)	2 (40)
C	2 (40)	1 (20)
D	1 (20)	1 (20)
Secondary HPF, n (%)	1 (20)	0
Mediastinal shift, n (%)		
Left	1 (20)	1 (20)
Right	2 (40)	2 (40)
None	1 (20)	0

EGA = estimated gestational age.
Percentages are expressed as column percentages.

Table 2
Patient management and outcomes.

	Non-survivors	Survivors
	n = 5	n = 5
Day of life intubated, n (%)		
0	2 (40)	3 (60)
1	0	1 (20)
12	1 (20)	0
Never intubated	0	1 (20)
ECMO utilized, n (%)	2 (40)	1 (20)
Repair completed, n (%)	5 (100)	4 (80)
Operated on day of life, median (range)	6 (2–20)	5.5 (4–13)
Separation of liver and lung, n (%)		
Complete	2 (40)	0
Partial	2 (40)	4 (100)
Not separated	1 (20)	0
Patch repair, n (%)	3 (60)	3 (60)
LOS in days, median (range)	29 (9–301)	33 (27–84)
Oxygen status at 30 days, n (%)[£]		
Ventilator or CPAP	2 (100)	2 (40)
Nasal cannula	0	1 (20)
Room air	0	3 (60)
Oxygen status at hospital discharge/transfer, n (%)[¶]		
Ventilator	2 (100)	0
Nasal cannula	0	3 (60)
Room air	0	3 (60)
Days from surgery until death, median (range)	21 (3–250)	N/A

ECMO = extracorporeal membrane oxygenation; LOS = length of stay prior to discharge, transfer, or death.
[£] Among the 7 patients who survived to 30 days.
[¶] Among the 7 patients who survived to discharge or transfer.

Table 3
Results of literature search.

First author	Year of publication	Title	Number of patients	Comments
Macpherson	1977	Pseudosequestration	3	1 Patient excluded (eventration)
Tsugawa	1987	Right congenital diaphragmatic hernia: anatomical significance and surgical repair	2	Excluded – not available in English
Katz	1998	Fibrous fusion between the liver and the lung: an unusual complication of right congenital diaphragmatic hernia	1	
Slovis	2000	Hepatic pulmonary fusion in neonates	6	
Gasmi	2003	Unusual discovery of hepato-pulmonary fusion during right congenital diaphragmatic hernia surgery	1	Excluded – not available in English
Keller	2003	MR imaging of hepatic pulmonary fusion in neonates	1	
Robertson	2006	Right congenital diaphragmatic hernia associated with fusion of the liver and the lung	1	
Tanaka	2006	Treatment of a case with right-sided diaphragmatic hernia associated with an abnormal vessel communication between a herniated liver and the right lung	1	
Gander	2010	Hepatic pulmonary fusion in an infant with a right-sided congenital diaphragmatic hernia and contralateral mediastinal shift	1	
Taide	2010	Hepatic pulmonary fusion: a rare case	1	
Khatwa	2010	Multidetector computed tomography evaluation of secondary hepatopulmonary fusion in a neonate	1	
Castle	2011	Right-sided congenital diaphragmatic hernia, hepatic pulmonary fusion, duodenal atresia, and imperforate anus in an infant	1	Excluded – not HPF
Chandrashekhara	2011	Hepatic pulmonary fusion: case report with review of literature	1	
Breysem	2012	Multidetector CT of right-sided congenital diaphragmatic hernia associated with hepatopulmonary fusion in a newborn	1	
Hamilton	2012	Fatal complication after repair of a congenital diaphragmatic hernia associated with hepatopulmonary fusion, anomalous right pulmonary venous return, and azygos continuation of the inferior vena cava	1	
Lin	2012	Hepatic pulmonary fusion: Two cases with diaphragmatic hernia and one case with Pentalogy of Cantrell	3	
Saurabh	2013	Hepatic pulmonary fusion: a rare association of right-sided congenital diaphragmatic hernia	1	
Olenik	2014	Hepatopulmonary fusion in a newborn. An uncommon intraoperatory finding during right congenital diaphragmatic hernia surgery: case description and review of literature	1	
Laamiri	2016	Right congenital diaphragmatic hernia associated with hepatic pulmonary fusion: A case report	1	
Jain	2017	Hepatopulmonary fusion: a rare and potentially lethal association with right congenital diaphragmatic hernia	1	
Takezoe	2017	Prenatally diagnosed, right-sided congenital diaphragmatic hernia complicated by hepatic pulmonary fusion and intrathoracic kidney	1	
Almaramhy	2018	Hepatopulmonary fusion associated with right-sided congenital diaphragmatic hernia: management of this rare anomaly and a review of the literature	1	
Kerkeni	2018	How to treat hepatic pulmonary fusion: case report with review of literature	1	

communicating between the liver and lung (Fig. 1C). The attempt to separate the organs was aborted and plication was performed instead, as described above. In 4 cases, separation of the medial aspect of the fusion was not attempted due to proximity to the pulmonary hilum and inferior vena cava (IVC). Instead, the liver and lung were left fused medially and the diaphragmatic defect was repaired around the fused structures.

One male infant underwent CDH repair on DOL 2 while on ECMO support, and HPF was not observed during the index operation. However, the patient underwent reoperation on DOL 14 due to persistent opacification of the right hemithorax and was found to have secondary HPF. Partial separation and liver reduction was achieved and patch repair was performed. The patient's postoperative course was complicated by abdominal compartment syndrome requiring emergent decompression and eventual multi-organ failure. The patient expired on postoperative day 6 after reoperation (DOL 23).

Of note, 2 survivors, including the patient who did not undergo CDH repair, underwent cardiac catheterization with embolization of large aorto-pulmonary collateral vessels. Both patients subsequently demonstrated improvement of pulmonary hypertension and overall respiratory status.

3. Discussion

Macpherson may have been the first author to describe HPF in 1977, although he termed the anomaly "pseudosequestration." [3] His series of 3 patients included 2 patients with right-sided CDH and lung that was adherent through the diaphragmatic defect. (The other patient had diaphragm eventration, excluding the diagnosis of HPF.) In order to achieve separation, 1 patient underwent partial hepatectomy and the other underwent right lower lobectomy, suggesting marked adhesion between the organs.

The first definite case of HPF, reported by Katz in 1998, described an inseparable fusion of liver and lung in a neonate with central CDH [4]. Since then, HPF has been described in multiple case reports and series [5]. An extensive review of the literature revealed 32 potential cases of HPF described in 23 different reports (Table 3). Of those, 2 papers describing HPF in 3 patients were not available for review in English, and 1 paper incorrectly labeled an adhesion between the peritoneum and right lung as HPF [6–8]. Thus, 28 previous cases were available for review, in addition to the 10 reported here (Online Supplement 1).

The prevalence of HPF within the CDHR is approximately 3 in 1000 infants (7 cases of HPF out of 2265 cases of CDH in the CDHR from January 2015 through June 2019). HPF appears to affect both sexes equally (Table 4). Overall mortality among reported cases is 49% ($n = 18$). However, it is difficult to determine how many deaths are attributable to HPF and how many are caused by the underlying CDH with associated pulmonary hypertension and hypoplasia.

The diagnosis of HPF is almost always made intraoperatively. Previous reviews have focused on the direction of mediastinal shift as a potential preoperative indicator of HPF, as a number of patients were observed to have rightward or absent mediastinal shift on radiographs [5,9]. Among all cases with known imaging results ($n = 30$), mediastinal shift was rightward in 30%, absent in 27%, and leftward in 43%; thus, plain films fail to warn the surgeon of the underlying anomaly in nearly half of affected patients. Advanced imaging modalities, such as CT or magnetic resonance imaging (MRI), are more likely to demonstrate HPF, particularly when intravenous contrast is used. One patient in our series was diagnosed by CT angiography but has never received

Table 4
All reported cases of hepatopulmonary fusion.

	Non-survivors (n = 18)		Survivors (n = 19)	
	n / n with data	%	n / n with data	%
Female	10/18	55.6%	8/18	44.4%
Born full term	13/14	92.9%	9/9	100.0%
Presented at ≥2 months	2/18	11.1%	6/19	31.6%
Advanced imaging performed[¶]	7/16	43.8%	10/15	66.7%
Cardiac anomalies	6/18	33.3%	4/18	22.2%
Vascular anomalies	9/18	50.0%	15/18	83.3%
Right pulmonary vascular anomalies	9/18	50.0%	13/18	72.2%
Aorta anomalies	2/17	11.8%	1/16	6.3%
IVC anomalies	4/17	23.5%	4/16	25.0%
Hepatic vascular anomalies	1/17	5.9%	4/16	25.0%
Other (non-cardiovascular) anomalies	6/17	35.3%	2/16	12.5%
Mediastinal shift				
None	4/14	28.6%	4/16	25.0%
Right	4/14	28.6%	5/16	31.3%
Left	6/14	42.9%	7/16	43.8%
Secondary HPF	2/18	11.1%	2/19	10.5%
ECMO utilized	3/18	16.7%	1/19	5.3%
Underwent operative repair	17/18	94.4%	18/19	94.7%
Operative timing				
Neonatal period	15/17	88.2%	12/18	66.7%
1 month to 1 year	2/17	11.8%	4/18	22.2%
>1 year	0/17	0.0%	2/18	11.1%
Patch repair	7/13	53.8%	7/9	77.8%
Degree of liver-lung separation achieved				
Not separated	2/15	13.3%	2/15	13.3%
Partial	6/15	40.0%	5/15	33.3%
Complete	7/15	46.7%	8/15	53.3%
Method of separating liver and lung[¥]				
Sharp	0/8	0.0%	1/9	11.1%
Cautery	2/8	25.0%	0/9	0.0%
LigaSure	3/8	37.5%	2/9	22.2%
Hepatic or pulmonary resection	3/8	37.5%	6/9	66.7%
Hepatic tissue resected	2/16	12.5%	2/14	14.3%
Pulmonary tissue resected	2/16	12.5%	5/14	35.7%
Days from birth to death, median (IQR)[µ]	10.5 (7–29)		N/A	
Days from surgery to death, median (IQR)[§]	4 (1.5–9.5)		N/A	

[¶] Includes CT/CTA, MRI/MRA, arteriography, venography.
[¥] For patients who underwent partial or complete separation of the liver and lung.
[µ] For the 10 patients with available data.
[§] For the 12 patients with available data.

surgical confirmation of the diagnosis. Chandrashekhara also reported a preoperative diagnosis of HPF by MRI [10]. However, preoperative contrast-enhanced CT has failed to establish the diagnosis in several cases [11–13].

Advanced imaging may also aid in the identification of abnormal vasculature, which is frequently reported in association with HPF. The frequency with which anomalous right pulmonary vasculature has been observed in HPF has led several authors to suggest a relationship with congenital pulmonary venolobar syndrome (Scimitar syndrome), a condition in which the right pulmonary veins drain into the IVC [10,11,14]. Scimitar syndrome has an estimated prevalence of 1 to 3 per 100,000 births, and infants presenting with scimitar syndrome are reported to have a relatively poor prognosis, with a 1-year survival rate of 55–63% [15]. Several cases of Scimitar syndrome associated with right-sided CDH, in the absence of HPF, have been reported [16–18]. However, it is unclear whether HPF and scimitar syndrome share a common etiology, as the pathogenesis of CDH, with or without HPF, remains as yet undefined.

Among reported cases of HPF, 60% of patients had aberrant right pulmonary vasculature (n = 22), including: systemic arterial supply, venous drainage into the liver or IVC, and hypoplastic or absent pulmonary arteries and/or veins. Hamilton described an infant who underwent complete

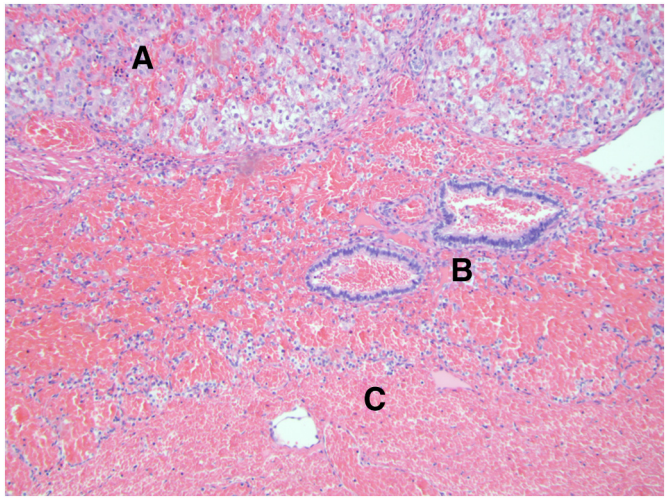

Fig. 2. Histopathology of hepatopulmonary fusion specimen showing hepatocytes (A), bronchi (B), and alveoli (C) without a plane of separation.

separation of the liver and lung but died shortly after surgery [19]. Autopsy revealed an absent intrahepatic IVC, infrahepatic systemic venous drainage into the superior vena cava, and anomalous drainage of the right pulmonary vein into the supradiaphragmatic IVC. The bowel was also noted to have severe vascular congestion. The patient's death was attributed to his underlying vascular anomalies, which were not identified prior to or during surgery.

Anomalous vasculature may contribute to some patients' poor outcomes, but it may also represent a target for intervention. Tanaka described a female neonate with HPF who demonstrated failure to thrive and inability to wean from the ventilator for months after plication of the right diaphragmatic hernia membrane [20]. Angiography performed at 1 year of age demonstrated right pulmonary arterial supply by an anomalous hepatic artery branch, right pulmonary venous drainage into the liver through aberrant vessels between the liver and right lung, and the absence of right pulmonary venous drainage into the left atrium. The anomalous vessels were ligated during the ensuing reoperation for CDH repair, and the patient improved postoperatively and was weaned from the ventilator. Two patients in our series similarly underwent cardiac catheterization with embolization of large aorto-pulmonary collateral vessels with subsequent improvement in their respiratory statuses. The success observed in these patients suggests that a thorough understanding of the patient's cardiovascular physiology is necessary to guide treatment decisions. Once the diagnosis of HPF is suspected or confirmed, vascular imaging should be obtained whenever possible. Involving a cardiologist may also be warranted.

In addition to the frequent presence of vascular anomalies, the absence of clear tissue planes makes separation of the liver and lung technically challenging (Fig. 2). The degree of fusion between liver and lung parenchyma is variable, ranging from “a delicate fibrous membrane,” to “partial continuity” of the parenchyma, to complete fusion with no identifiable tissue planes [5,21,22]. Numerous surgical approaches have been described. Surgeons have achieved complete separation of the organs using electrocautery, tissue resection (partial hepatectomy or pulmonary lobectomy), or sharp dissection [3,11,13,14,16,19,21,23]. Tanaka reported a staged approach, in which plication of the hernia membrane was performed during the index operation. When the patient's respiratory status failed to improve postoperatively, the patient underwent a second operation at 13 months, in which a partial hepatectomy of segments 6 and 7 was performed [20]. Other surgeons have avoided complete separation of the organs [4,9,10,22,24–27]. Gander reported using polyester fiber mesh on the liver capsule and approximating the diaphragmatic defect around the HPF without attempting separation [24]. Robertson described leaving the medial

portion of the lung fused to the liver and closing the majority of the diaphragm defect around the HPF [26]. A similar approach was utilized for several patients in this series.

Secondary HPF, organ fusion observed during reoperation but not during the index case, has been previously reported in 3 patients and was noted in 1 patient in our series [12,13,24]. Takezoe reported that a biopsy taken of the posterior diaphragm of 1 of these patients during the index operation demonstrated nests of Hep-1 positive hepatocytes and keratin-7 positive bile duct cells without any detectable muscle tissue. These histopathologic findings suggest a congenital predisposition for the development of secondary HPF. However, it is unclear whether primary and secondary HPF share the same etiology. When feasible, surgeons should consider obtaining a biopsy of the diaphragm in affected patients, as this may yield further insight into the etiology of HPF.

4. Conclusions

This case series and literature review elicit several important observations. Preoperative diagnosis is generally not possible, though absent or rightward mediastinal shift may provide a clue (as leftward shift is expected in right-sided CDH). Moreover, the majority of HPF patients have underlying vascular anomalies, which may be identifiable on preoperative imaging, and these anomalies may contribute to serious intraoperative or postoperative complications. Diagnostic and therapeutic catheterization may be useful in some patients. While several operative approaches have been described, complete separation may not be ideal, given the frequency of vascular anomalies. Partial separation laterally, leaving the medial fusion intact, in combination with a generous diaphragmatic patch may be the optimal approach. Finally, survival is only slightly lower than the registry overall, though lower birthweight and ECMO support portend a poorer prognosis. Knowledge about this rare entity among CDH patients should inform family counseling and clinical decision-making.

Appendix A. Contributing Authors

Institution	Contributing Author
Akron Children's Hospital	Jo Ann Lindeman, APRN, CNP
Duke University	Kimberly A. Fisher, PhD
Johns Hopkins All Children's Hospital	Dina Fouad, MBChB David W. Kays, MD
Karolinska Institutet	Carmen Mesas Burgos, MD, PhD
Polish Mother's Health Center Research Institute	Iwona A. Maroszyńska, MD, PhD
Pontificia Universidad Catolica de Chile	Matias Luco, MD
Saint Louis University	Melissa J. Hawkins, MSN, RN, CPNP Gustavo A. Villalona, MD
University of Alabama at Birmingham and Children's of Alabama	Robert T. Russel, MD Leah J. Schoel, BA
University of North Carolina at Chapel Hill School of Medicine	Sean E. McLean, MD
University of Texas John P. and Katherine G. McGovern Medical School	Dalya M. Ferguson, MD Matthew T. Harting, MD, MS Kevin P. Lally, MD, MS Pamela A. Lally, MD KuoJen Tsao, MD
University of Virginia	Daniel E. Levin, MD Eugene D. McGahren, MD

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2019.09.037.

References

[1] Taide D, Bendre P, Kirtane J, et al. Hepatic pulmonary fusion – A rare case. Afr J Paediatr Surg 2010;7:28–9.
[2] Tsao KJ, Lally KP. The Congenital Diaphragmatic Hernia Study Group: a voluntary international registry. Semin Pediatr Surg 2008;17:90–7. https://doi.org/10.1053/j.sempedsurg.2008.02.004.
[3] Macpherson RI, Whytehead L. Pseudosequestration. J Can Assoc Radiol 1977;28:17–25.
[4] Katz S, Kidron D, Litmanovitz I, et al. Fibrous fusion between the liver and the lung: An unusual complication of right congenital diaphragmatic hernia. J Pediatr Surg 1998;33:766–7. https://doi.org/10.1016/S0022-3468(98)90214-7.
[5] Slovis TL, Farmer DL, Berdon WE, et al. Hepatic pulmonary fusion in neonates. Am J Roentgenol 2000;174:229–33. https://doi.org/10.2214/ajr.174.1.1740229.
[6] Gasmi M, Mekki M, Jouini R, et al. Unusual discovery of hepato-pulmonary fusion during right congenital diaphragmatic hernia surgery. Presse Med 2003;32:460–1.
[7] Tsugawa C, Matsumoto Y, Sugimura C, et al. Right congenital diaphragmatic hernia: anatomical significance and surgical repair. Jpn J Pediatr Surg 1987;19:931–5.
[8] Castle SL, Naik-Mathuria BJ, Torres MB. Right-sided congenital diaphragmatic hernia, hepatic pulmonary fusion, duodenal atresia, and imperforate anus in an infant. J Pediatr Surg 2011;46:1432–4. https://doi.org/10.1016/j.jpedsurg.2011.01.024.
[9] Almaramhy HH. Hepatopulmonary fusion associated with right-sided congenital diaphragmatic hernia: management of this rare anomaly and a review of the literature. J Int Med Res 2018;030006051875989. https://doi.org/10.1177/0300060518759892.
[10] Chandrashekhara SH, seith Bhalla A, Gupta AK, et al. Hepatic pulmonary fusion: case report with review of literature. J Pediatr Surg 2011;46:e23–7. https://doi.org/10.1016/j.jpedsurg.2010.11.032.
[11] Breysem L, Vanhaesebrouck S, Gewillig M, et al. Multidetector CT of right-sided congenital diaphragmatic hernia associated with hepatopulmonary fusion in a newborn. Pediatr Radiol 2012;42:1138–41. https://doi.org/10.1007/s00247-012-2379-1.
[12] Khatwa U, Lee EY. Multidetector computed tomography evaluation of secondary hepatopulmonary fusion in a neonate. Clin Imaging 2010;34:234–8. https://doi.org/10.1016/j.clinimag.2009.07.005.
[13] Takezoe T, Nomura M, Ogawa K, et al. Prenatally diagnosed, right-sided congenital diaphragmatic hernia complicated by hepatic pulmonary fusion and intrathoracic kidney. Birth Defects 2017;1. https://doi.org/10.15761/BDJ.1000104.
[14] Jain V, Yadav DK, Kandasamy D, et al. Hepatopulmonary fusion: a rare and potentially lethal association with right congenital diaphragmatic hernia. BMJ Case Rep 2017;2017:bcr2016218227. https://doi.org/10.1136/bcr-2016-218,227.
[15] Wang H, Kalfa D, Rosenbaum MS, et al. Scimitar syndrome in children and adults: natural history, outcomes, and risk analysis. Ann Thorac Surg 2018;105:592–8. https://doi.org/10.1016/J.ATHORACSUR.2017.06.061.
[16] Kerkeni Y, Farhani R, Sassi N, et al. How to treat hepatic pulmonary fusion: case report with review of literature. Acta Chir Belg 2018:1–3. https://doi.org/10.1080/00015458.2018.1496568.
[17] Rossetti LZ, Glinton K, Yuan B, Liu P, Pillai N, Mizerik E, et al. Review of the phenotypic spectrum associated with haploinsufficiency of MYRF. Am J Med Genet 2019;179:ajmg.a.61182. doi:https://doi.org/10.1002/ajmg.a.61182.
[18] Youssef T, Mahmoud H, Ionescu S, et al. Scimitar syndrome associated with aberrant right subclavian artery, diaphragmatic hernia, and urinary anomalies-case report and review of the literature. Romanian J Morphol Embryol 2018;59:625–30.
[19] Hamilton J, Jaroszewski D, Notrica D. Fatal complication after repair of a congenital diaphragmatic hernia associated with hepatopulmonary fusion, anomalous right pulmonary venous return, and azygos continuation of the inferior vena cava. Eur J Pediatr Surg 2012;24:350–2. https://doi.org/10.1055/s-0032-1,324,695.
[20] Tanaka S, Kubota M, Yagi M, et al. Treatment of a case with right-sided diaphragmatic hernia associated with an abnormal vessel communication between a herniated liver and the right lung. J Pediatr Surg 2006;41:e25–8. https://doi.org/10.1016/j.jpedsurg.2005.12.032.
[21] Olenik D, Codrich D, Gobbo F, et al. Hepatopulmonary fusion in a newborn. An uncommon intraoperatory finding during right congenital diaphragmatic hernia surgery: case description and review of literature. Hernia 2014;18:417–21. https://doi.org/10.1007/s10029-012-1042-y.
[22] Laamiri R, Belhassen S, Ksia A, et al. Right congenital diaphragmatic hernia associated with hepatic pulmonary fusion: A case report. J Neonatal Surg 2016;5:35. https://doi.org/10.21699/jns.v5i3.370.
[23] Lin J, Durham MM, Ricketts R, et al. Hepatic pulmonary fusion: two cases with diaphragmatic hernia and one case with Pentalogy of Cantrell. Fetal Pediatr Pathol 2012;31:401–9. https://doi.org/10.3109/15513815.2012.659406.
[24] Gander JW, Kadenhe-Chiweshe A, Fisher JC, et al. Hepatic pulmonary fusion in an infant with a right-sided congenital diaphragmatic hernia and contralateral mediastinal shift. J Pediatr Surg 2010;45:265–8. https://doi.org/10.1016/j.jpedsurg.2009.10.090.
[25] Keller RL, Aaroz PA, Hawgood S, et al. MR imaging of hepatic pulmonary fusion in neonates. Am J Roentgenol 2003;180:438–40. https://doi.org/10.2214/ajr.180.2.1800438.
[26] Robertson DJ, Harmon CM, Goldberg S. Right congenital diaphragmatic hernia associated with fusion of the liver and the lung. J Pediatr Surg 2006;41:e9–10. https://doi.org/10.1016/j.jpedsurg.2006.02.031.
[27] Saurabh K, Kumar S, Chellani H, et al. Hepatic pulmonary fusion: a rare association of right-sided congenital diaphragmatic hernia. Ann Gastroenterol 2013;26:95–6.