

Case Report

A case series of right-sided congenital diaphragmatic hernia with liver herniation and non-contralateral mediastinal shift: Imaging clues and the diagnostic challenge of hepatopulmonary fusion

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Abstract

Right-sided congenital diaphragmatic hernia (CDH) with liver herniation typically produces contralateral mediastinal shift; its absence presents a diagnostic imaging challenge and raises concern for hepatopulmonary fusion (HPF). We report three infants with right-sided CDH and liver herniation non-contralateral mediastinal shift, including two cases of HPF and one non-HPF case attributable to severe pulmonary hypoplasia. This series demonstrates that non-contralateral mediastinal shift, although classically associated with HPF, is not pathognomonic. We highlight the complementary roles and limitations of radiography, ultrasonography, and contrast-enhanced CT in suggesting HPF preoperatively. These cases refine the radiologic clues for this rare presentation and underscore the importance of meticulous multimodality imaging to inform surgical planning and anticipate operative complexity.

Keywords: Hepatopulmonary fusion, Non-contralateral mediastinal shift, Right-sided congenital diaphragmatic hernia.

Introduction

Congenital diaphragmatic hernia (CDH) is a rare congenital anomaly, occurring in approximately 2.4–4.2 per 10,000 births, and results from the failure of fusion between the transverse septum and pleuroperitoneal folds during the 8th–10th weeks of gestation, leading to herniation of abdominal viscera into the thoracic cavity and subsequent pulmonary hypoplasia [1–4]. The majority of CDH cases are posterolateral (Bochdalek) defects and are predominantly left-sided, whereas right-sided and bilateral defects are less common [1,5–7]. In right-sided CDH with liver herniation, the mass effect typically produces a contralateral mediastinal shift; however, rare cases with non-contralateral mediastinal shift pose a diagnostic challenge and raise the possibility of severe right lung hypoplasia or hepatopulmonary fusion (HPF) [8]. HPF is an exceptionally uncommon anomaly, reported in approximately 3 in 1,000 right-sided CDH cases, characterized by abnormal fusion between the herniated liver and right lung and frequently associated with aberrant pulmonary vasculature, which increases surgical complexity and adversely affects outcomes [9,10]. Pre-operative imaging often fails to definitively identify HPF, and many cases are diagnosed intraoperatively. However, careful multimodality imaging, particularly through detailed evaluation of anatomic and vascular relationships, may facilitate preoperative recognition and guide surgical planning [6,11,12]. Between 2014 and 2025, our institution encountered three cases of right-sided CDH with liver herniation and non-contralateral mediastinal shift, including two with HPF and one without, which form the basis of this report, emphasizing the imaging approaches and findings relevant to the diagnosis.

Case summary

Case 1

A term male newborn delivered by cesarean section for breech presentation developed respiratory distress at birth, requiring intubation. Initial chest-abdominal radiographs showed near-complete opacification of the right hemithorax with air-filled bowel loops and a mild contralateral mediastinal shift, suggesting right-sided CDH with liver and bowel herniation (Figure 1A). A follow-up radiograph further supported the diagnosis, demonstrating a well-defined elevated hepatic contour within the thorax, an abnormally high right hemidiaphragm, the absence of visible herniated bowel loops, and non-contralateral mediastinal shift (Figure 1B).

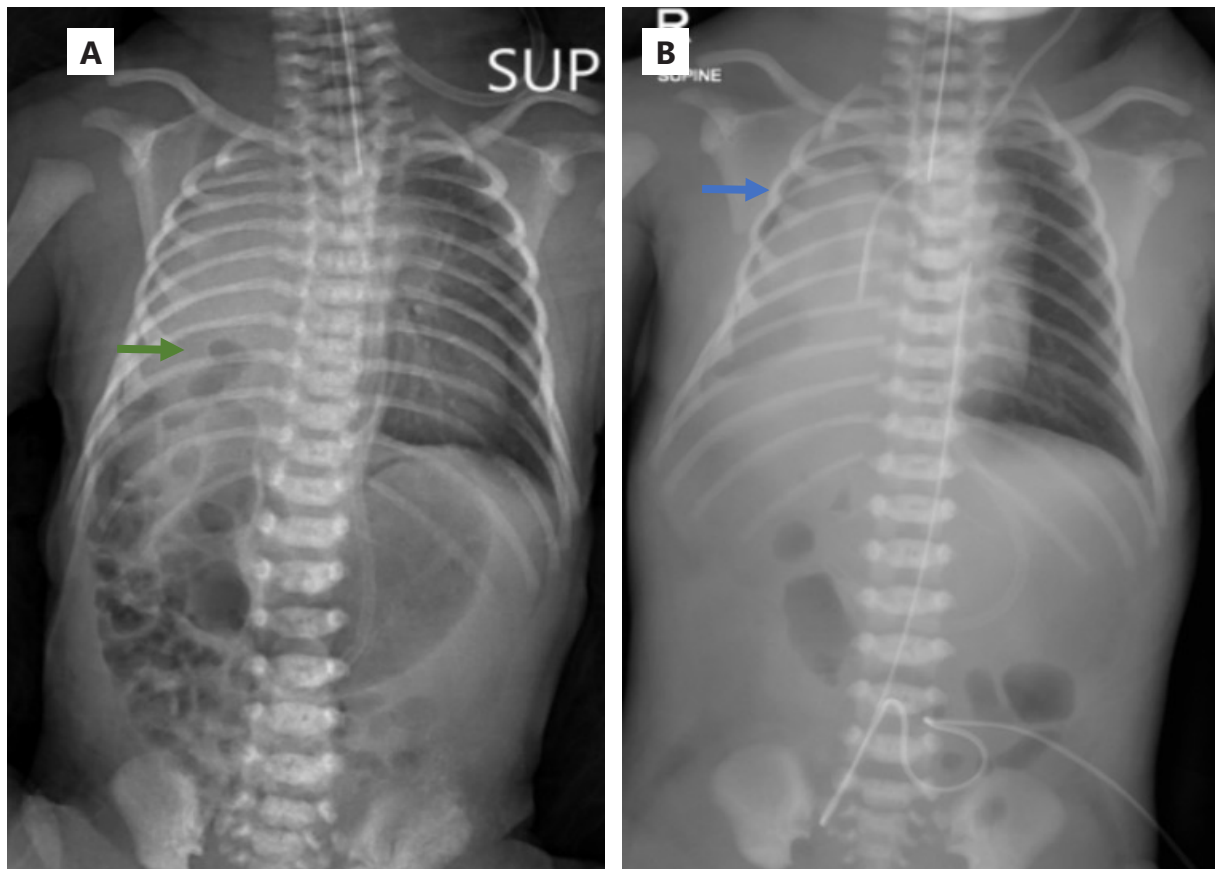


Figure 1. Chest-abdominal radiographs of a term newborn with dyspnea: (A) Day of life (DOL) 1, the radiograph showed near-total right hemithoracic opacification with air-filled bowel loops (green arrow) and a mild contralateral mediastinal shift, consistent with liver and bowel herniation. (B) On DOL 2, the follow-up radiograph showed a sharply elevated hepatic contour (blue arrow) with non-contralateral mediastinal shift and minimal to no visible herniated bowel loops.

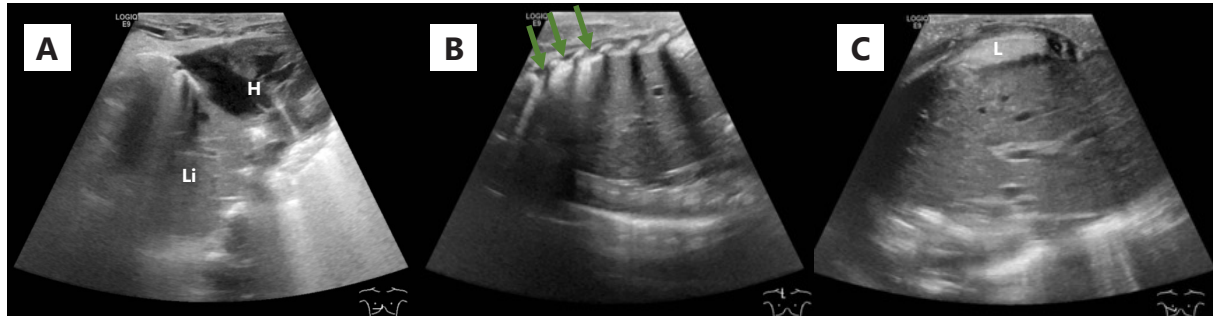


Figure 2. *US demonstrated an abnormally elevated right hepatic lobe within the right hemithorax, directly abutting the aerated right lung and heart, with no identifiable right hemidiaphragm (A). Aerated lung tissue (green arrows) was seen interposed between two portions of the herniated liver (B). The liver was seen in close contact with the atelectatic right lung within the hemithorax (C).*
Li = liver; H = heart; L = lung.

Chest ultrasonography (US) confirmed intrathoracic displacement of the right hepatic lobe with absence of the right hemidiaphragm, partial interposition of aerated right lung, and close apposition of hepatic tissue to the non-aerated lung. Hepatic venous drainage into the inferior vena cava (IVC) was preserved (Figure 2).

Intraoperatively, a right hemidiaphragmatic defect with herniation of the right hepatic lobe and fusion to hypoplastic right lung tissue was observed. Partial repair with a patch was performed on DOL 15.

At 10 months, CT angiography demonstrated an atelectatic right lung in direct contact with and inseparable from the herniated right hepatic tissue. The right pulmonary artery was hypoplastic, and the right pulmonary vein was absent (Figure 3). The right hemidiaphragm remained absent, while the portal and hepatic veins drained normally into the IVC.

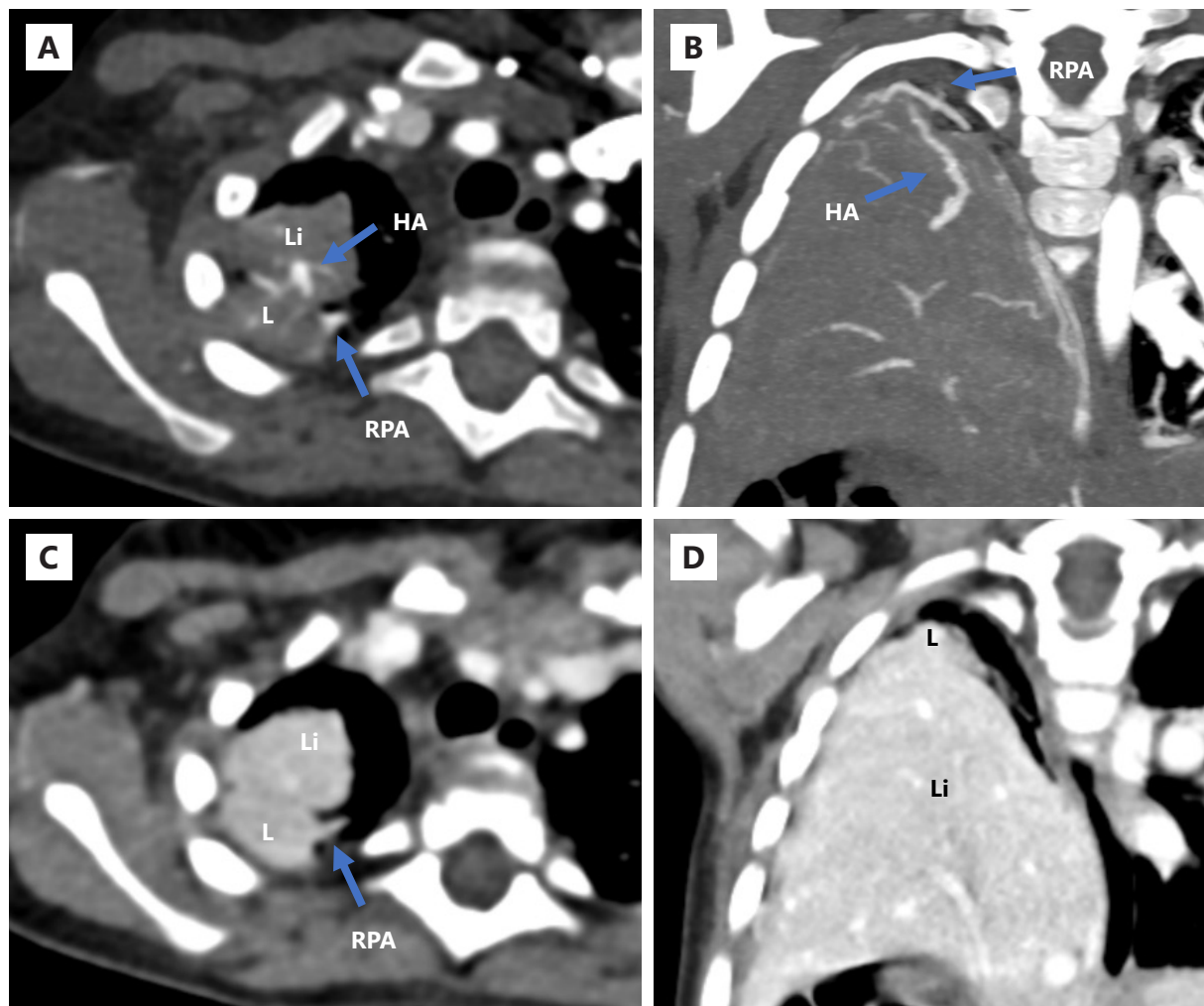


Figure 3. CT angiography of a 10-month-old infant with right-sided CDH and HPF, status post partial repair of the diaphragmatic defect on DOL 15, showed an enhancing atelectatic right lung conforming to the dome of the herniated right hepatic lobe within the right hemithorax on both the arterial (A,B) and venous (C,D) phases. The lung parenchyma received its vascular supply from a branch of the hypoplastic right pulmonary artery.

L = lung; *Li* = liver; *RPA* = right pulmonary artery; *HA* = hepatic artery.

Persistent elevation of the right hemidiaphragm with right-sided opacity representing the herniated hepatic lobe and HPF remained evident on the follow-up radiographs. The patient has experienced recurrent episodes of pneumonia and bronchitis and requires home oxygen therapy.

Case 2

A term male newborn presented with tachypnea after birth. Chest radiographs demonstrated a lobulated contour with elevation of the right hemidiaphragm, suspicious for liver herniation, with adjacent pulmonary opacification and a non-contralateral mediastinal shift (Figure 4A).

Preoperative chest CT performed on the same day demonstrated marked elevation of the right hemidiaphragm with superior herniation of the right hepatic lobe into the right hemithorax, abutting a hypoplastic right lung supplied by a diminutive right pulmonary artery. A suspected anomalous pulmonary venous channel draining via the hepatic vein into the right atrium was noted (Figure 4B-D). A right-sided aortic arch with an aberrant left subclavian artery and a Kommerell diverticulum was also observed.

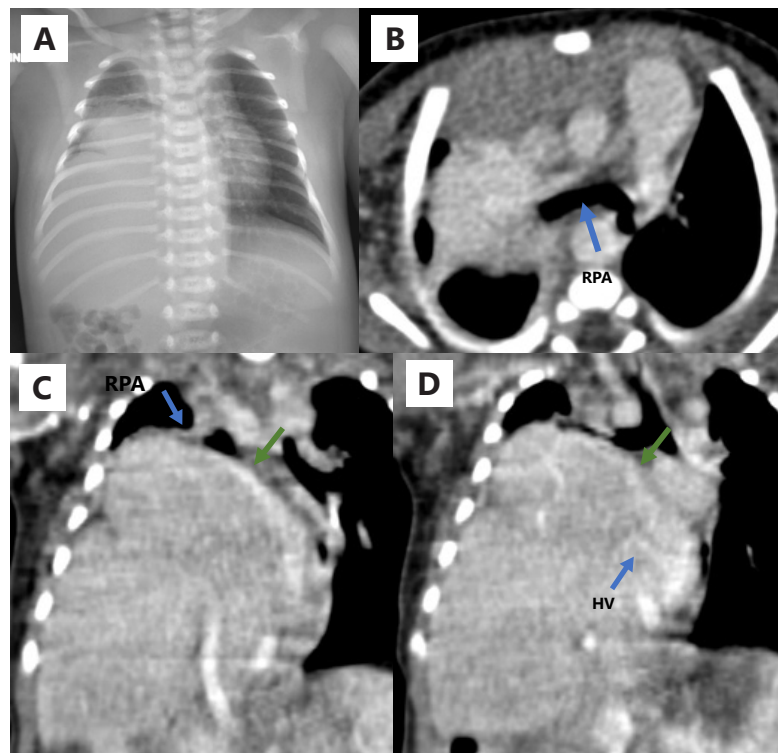


Figure 4. Preoperative chest radiograph (A) of a term male newborn with tachypnea after birth showed a lobulated contour and elevation of the right hemidiaphragm with adjacent pulmonary opacification, a small aerated portion of the right upper lobe, and a non-contralateral mediastinal shift. Preoperative chest CT (B-D) revealed marked elevation of the right hemidiaphragm with the herniated right hepatic lobe extending into the right hemithorax. The right lung was hypoplastic and supplied by a hypoplastic right pulmonary artery (RPA). A suspected anomalous pulmonary venous channel (green arrow) joined the hepatic vein (HV) and drained into the right atrium.

A diagnosis of right-sided CDH was made, and surgical repair was performed on the following day. Intraoperative findings confirmed right-sided CDH with HPF, evidenced by a right posterolateral diaphragmatic defect with herniation of the liver adherent to the right lower lobe pulmonary tissue. An arterial branch supplying the right lower lung was visualized; however, its origin was indeterminate. There was absence of the IVC with azygos continuation. The suprahepatic IVC was formed by the confluence of hepatic veins and an anomalous right pulmonary vein. Surgical management included separation of the fused lung and liver tissues followed by patch repair of the diaphragmatic defect (Figure 5A,B). Postoperative chest CT showed a clear distinction between the surgically separated pulmonary and hepatic tissues on the venous phase due to their different degrees of enhancement, whereas their similar enhancement of the tissues in certain portions on the arterial phase resulted in the partial inseparability (Figure 5C-F). Additional vascular anomalies were identified on postoperative CT, including a small right pulmonary vein draining into the left atrium and pleuropulmonary collateral vessels supplying the right basal lung from the right inferior phrenic artery (Figure 5G).

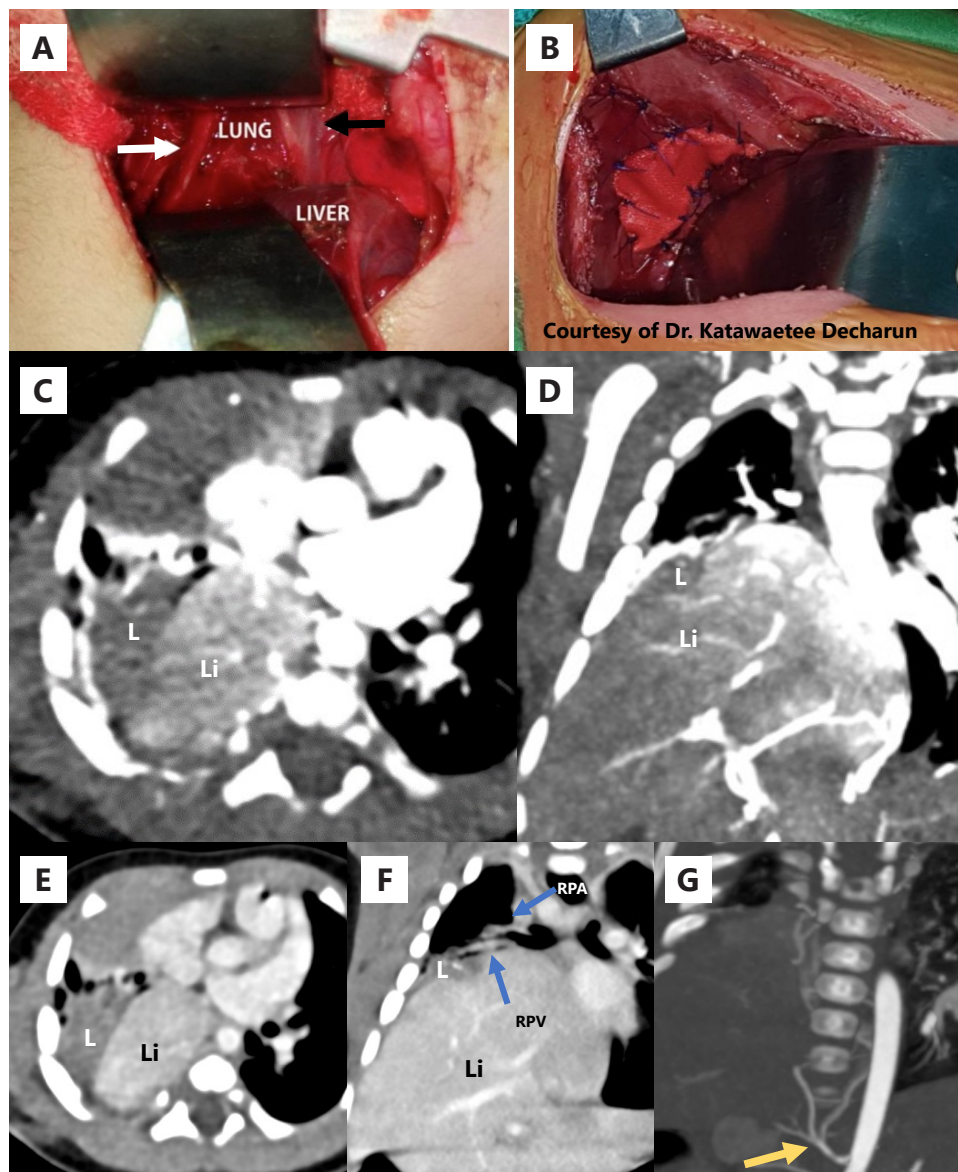


Figure 5. Intraoperative photographs (A,B) demonstrated a medially positioned arterial branch (white arrow) supplying the right lower lung, although its origin could not be identified. A draining vein (black arrow) was also observed. Patch repair of the diaphragmatic defect was performed following separation of the lung and liver tissues. Postoperative chest CT showed a partially indistinct interface between the surgically separated pulmonary and hepatic tissues on the arterial phase due to their similar enhancement (C,D). On the venous phase (E,F), the tissues were better delineated owing to the differential enhancement. Pleuropulmonary collateral vessels supplying the right basal lung from the right inferior phrenic artery were seen (G, yellow arrow). Li = liver; L = lung; RPA = right pulmonary artery; RPV = right pulmonary vein.

Postoperative chest radiographs continued to show elevation of the right hemidiaphragm, and subsequent ultrasonography confirmed right diaphragmatic paralysis (Figure 6). The patient has remained clinically stable, with only a single episode of pneumonia and reactive airway symptoms since surgery.

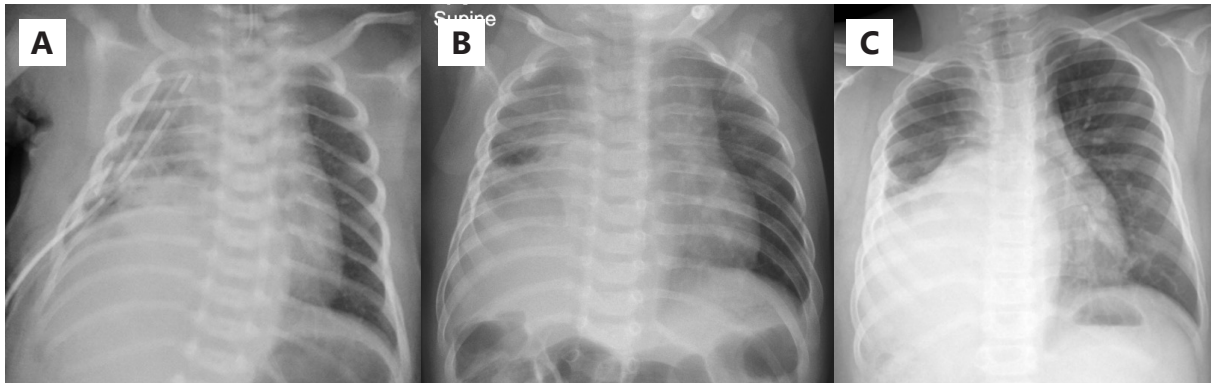


Figure 6. Chest radiograph obtained a few hours after surgery (A) showed an elevated right hemidiaphragm with right lung opacity and right pleural effusion following placement of an intercostal drainage catheter placement, consistent with postoperative changes. Follow-up radiographs at 3 months (B) and 6 years (C) demonstrated a persistently elevated right hemidiaphragm with a slight interval increase in the volume of the hypoplastic aerated right lung. Patchy opacities in the right lower lung zone likely represented atelectasis and possible pleural effusion. Right diaphragmatic paralysis was subsequently confirmed on ultrasound.

Case 3

A 4-month-old male infant with right-sided CDH and multiple congenital anomalies including global developmental delay, right hemifacial microsomia, right ear deformity, cleft lip, atrial septal defect, and anorectal malformation with rectourethral fistula status post colostomy, was referred to our institute for further surgical management.

Chest radiograph demonstrated right hemithoracic opacification with a non-contralateral mediastinal shift (Figure 7A). US showed herniation of the liver into the right thoracic cavity. An acute angulation between the intraabdominal and intrathoracic portions of the liver, the "waist sign", suggested a large diaphragmatic defect. No aerated right lung was identified, raising suspicion for right lung agenesis or marked hypoplasia with atelectasis (Figure 7B,C).

Chest CT at 6 months confirmed herniation of the right hepatic lobe into the right hemithorax, with only a small volume of atelectatic lung at the apex supplied by a hypoplastic right pulmonary artery (Figure 8).

Intraoperative findings revealed a posterior diaphragmatic defect involving nearly the entire right hemidiaphragm with herniation of the liver, which remained covered by a sac, into the right thoracic cavity. The herniated liver was successfully reduced into the abdominal cavity. The sac was opened, partially revised, and closed, followed by repair of the right diaphragmatic defect.

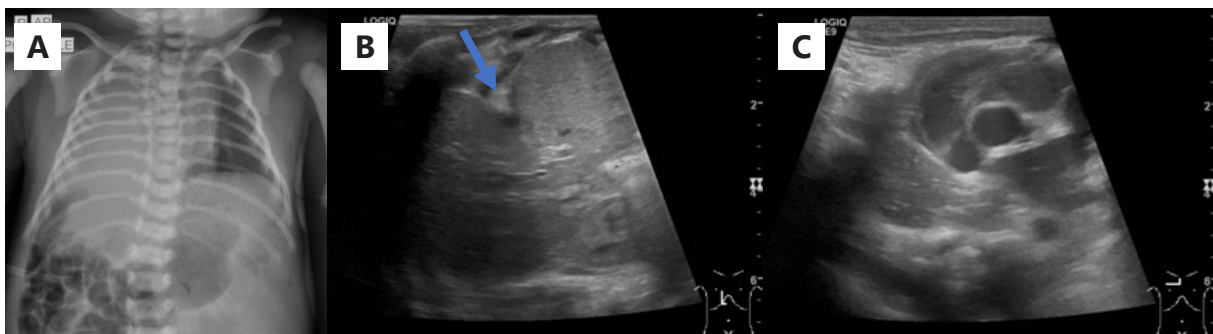


Figure 7. Chest radiograph showed right hemithoracic opacification with cardiac dextroposition (A). Ultrasound demonstrated liver herniation into the right thoracic cavity with acute angulation between the intraabdominal and intrathoracic portions of the liver or 'waist sign' (arrow), and no visible aerated right lung (B). The heart was located within the right hemithorax (C).

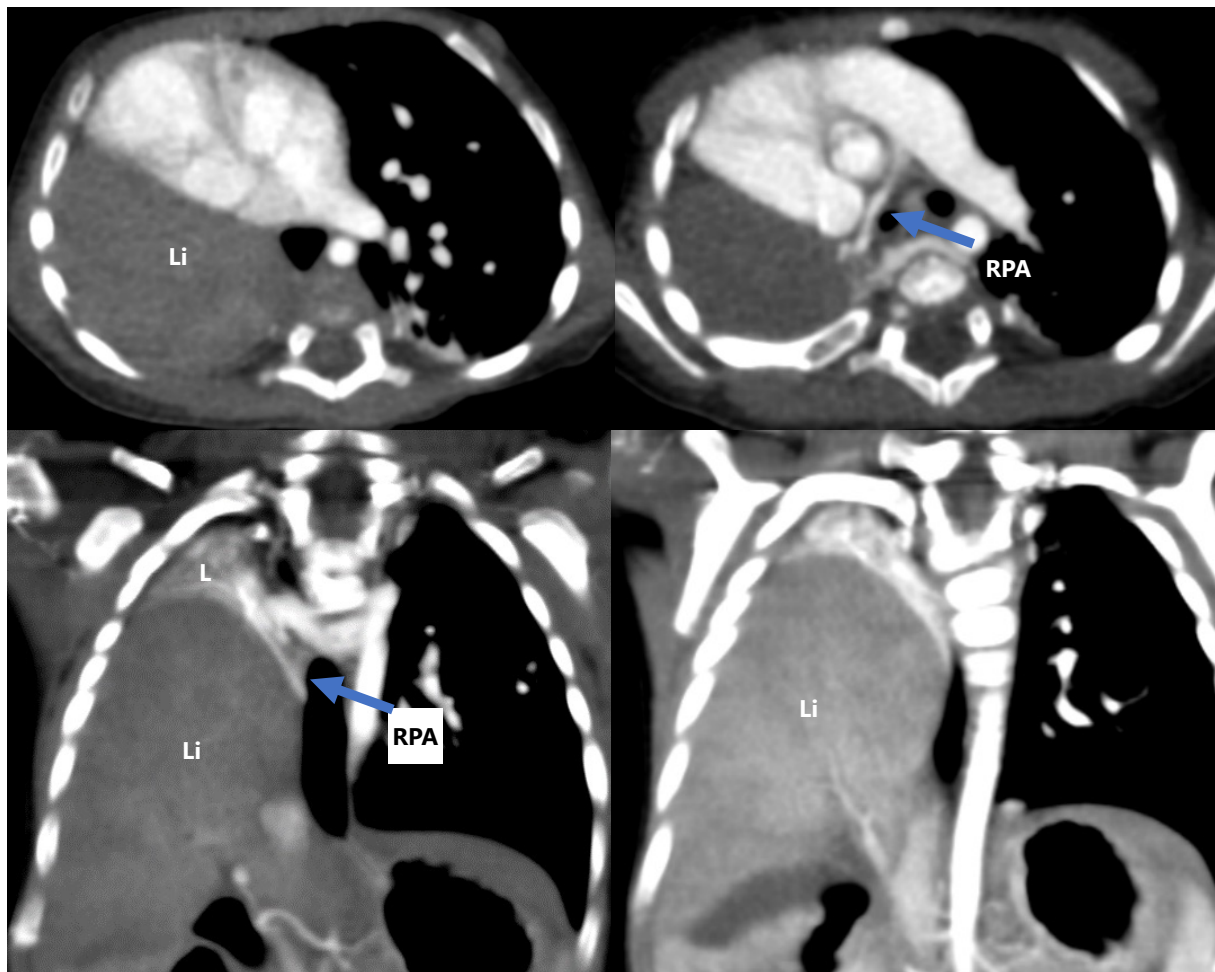


Figure 8. Chest CT at 6 months showed superior herniation of the right hepatic lobe (Li) into the right hemithorax, posterior to the heart. A small, compressed, atelectatic right lung remnant (L) was seen superiorly and was supplied by a hypoplastic right pulmonary artery (RPA).

A postoperative chest radiograph consistently showed persistent volume loss of the right hemithorax with compensatory hyperinflation of the left lung, consistent with a severe degree of right lung hypoplasia. The patient was discharged with home oxygen via nasal cannula. At the most recent follow-up, at age 7 months, he demonstrated adequate weight gain, clear sputum, and intermittent cyanosis. No further clinical information was available due to loss to follow-up.

Discussion

Imaging approaches for the rare presentation of right-sided CDH with liver herniation and non-contralateral mediastinal shift as well as the recognition of preoperative imaging features suggestive of HPF are challenging but can be valuable for operative planning and management, with important implications for patient outcomes.

Initial chest radiographs in our cases demonstrated typical features of right-sided CDH, including right hemithoracic opacification with tubular lucencies representing herniated bowel loops. These findings support chest radiography as an essential first-line modality for evaluating CDH and neonatal respiratory distress. Large or near-total opacification of the right hemithorax, often with an unusually elevated, curved right hemidiaphragm, raises suspicion for liver herniation and a relatively large diaphragmatic defect.

It has been recognized that right-sided CDH with liver herniation typically produces a contralateral mediastinal shift due to its mass effect. Non-contralateral mediastinal shift is therefore unusual and has prompted consideration of HPF [13], as in our cases. Although many reported HPF cases demonstrated non-contralateral mediastinal shift, following literatures indicated variability in this finding. A 2010 report described a case of right-sided CDH with HPF presenting with contralateral mediastinal shift, attributed to the size of the diaphragmatic defect and the degree of bowel herniation [14]. Concordantly, in our Case 1, the mild contralateral shift on day 1, when air-filled bowel loops were present, evolved to a non-contralateral shift on day 2 with persistent liver herniation and no visible bowel. In addition, a 2019 review found HPF cases to be evenly distributed between non-contralateral and contralateral mediastinal shift [9]. One of our cases was non-HPF at surgery, indicating that non-contralateral mediastinal shift in right-sided CDH with liver herniation is not specific for HPF and may reflect severe lung hypoplasia. Nonetheless, HPF should be considered in all cases of right-sided CDH [15] and should be strongly suspected when this atypical imaging pattern is present, highlighting the value of radiography in early recognition and in guiding further imaging.

US is a readily available adjunct for evaluating herniated organs and diaphragmatic integrity. In Case 3, the "waist sign," confirmed right-sided CDH with liver herniation into the right hemithorax; however, no right lung was visualized, limiting assessment for HPF and suggesting severe hypoplasia or agenesis. In contrast, Case 1 showed the right hepatic lobe herniating into the thorax, abutting the aerated lung and heart, with an absent right hemidiaphragm and aerated lung tissue interposed between the

hepatic portions - findings compatible with previously reported US findings of HPF [16]. These observations emphasize the value of US in diagnosing right-sided CDH and suggesting HPF.

Given the variability of mediastinal shift in HPF, this feature alone has limited diagnostic value [9]. Additional clues, particularly anomalous hepatopulmonary vasculature, are essential for raising suspicion for HPF. While US may show hepatic veins and IVC drainage, assessment is often limited. Contrast-enhanced CT is better suited for comprehensive evaluation.

Previously reported CT findings of HPF include anomalous right pulmonary vasculature and close contact at the lung-liver interface. However, details regarding enhancement patterns and imaging phases remain sparse. As a result, no definitive CT criteria have been established for the reliable diagnosis of HPF [9, 17]. In our HPF cases, CT demonstrated an atelectatic lung closely conforming to and inseparable from the herniated liver, with similar enhancement, as most evident on the venous phase. After surgical separation, the hepatic and pulmonary tissues showed distinct venous-phase enhancement, whereas arterial-phase similarity rendered them partially indistinguishable. In contrast, the non-HPF case showed a well-defined interface between the liver and the atelectatic lung. These observations support the concept that HPF may be suggested by inseparable hepatic and pulmonary tissues with similar enhancement, particularly on venous-phase CT, although caution is warranted given the small sample size and the nonuniform imaging protocols.

Additional CT features of HPF included right lung hypoplasia with a small right pulmonary artery and non-visualization of the right hemidiaphragm. CT suggested anomalous venous drainage in Case 2 was confirmed intraoperatively. Although small vessels and the origin of the aberrant systemic arterial supply to the right lower lung were not fully characterized, CT provided crucial anatomic information. While intraoperative confirmation remains the most reliable diagnostic method [9], the imaging clues and proposed features identified in our study highlight the value of preoperative CT in suggesting HPF and guiding surgical planning.

MRI may provide additional value in suspected HPF by demonstrating the loss of the normal interface between hepatic and pulmonary tissues and by delineating anomalous venous drainage; however, its role remains limited by the small number of reported cases and the lack of established diagnostic criteria [8, 11, 16, 18].

Postoperative radiographs showed persistent right hemithoracic opacity and hemidiaphragmatic elevation, reflecting lung hypoplasia, atelectasis, diaphragmatic paralysis, or postoperative changes related to partial diaphragmatic repair with residual HPF tissue. These findings underscore the importance of clinicoradiologic correlation.

Limitations include the small number of cases inherent to this rare condition, the retrospective nature of the image review, inconsistent CT protocols, and the lack of assessment of the role of MRI. Further studies with larger cohorts, standardized imaging protocols, and the incorporation of MRI findings, would be valuable for validating these observations and establish reliable imaging clues for distinguishing HPF from non-HPF entities.

Conclusion

Right-sided CDH with liver herniation and non-contralateral mediastinal shift is uncommon and has been recognized as a typical scenario for HPF; however, our series demonstrates that this pattern is not specific and may reflect severe pulmonary hypoplasia. Radiography, ultrasound, and CT are valuable for diagnosis and evaluation, and HPF should be considered in all such cases regardless of mediastinal shift, with non-contralateral mediastinal shift warranting heightened suspicion. Ultrasound may provide supportive findings, while CT is particularly useful for assessing anatomical relationships and detecting anomalous hepatopulmonary vasculature, although no definitive preoperative CT criteria exist. Our findings suggest that a conforming or inseparable liver-lung interface on contrast-enhanced CT may be a potential clue to HPF, but this observation requires further validation. At present, intraoperative assessment remains the most reliable diagnostic method, and the integration of clinical context with imaging findings is essential for optimal management.

Conflict of interest

The authors declare no conflict of interest.

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