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Horseshoe Lung Associated with Left Lung Hypoplasia: Case Report and Systematic Review of the Literature

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Summary

Horseshoe lung (HL) is often associated with cardiovascular malformations such as scimitar syndrome and unilateral lung hypoplasia. In patients with HL, the hypoplastic lung is almost always located on the right side. Cases of HL with a hypoplastic left lung are extremely rare.

In this paper, we describe a case of a one-day-old boy with HL involving left lung hypoplasia and perform a systematic review of the literature on HL with left lung hypoplasia.

Only 10 cases of HL involving left lung hypoplasia have been reported in the literature. Most of those cases also exhibited cardiovascular malformations and pulmonary hypertension. There have not been any reported cases of HL involving left lung hypoplasia associated with scimitar syndrome.

HL involving left lung hypoplasia is rare and tends to be associated with pulmonary hypertension.

MeSH Keywords: Cardiac Imaging Techniques • Lung • Scimitar Syndrome

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Background

Horseshoe lung (HL) is a rare congenital anomaly in which the right and left lungs are fused due to the formation of a narrow isthmus of lung parenchymal tissue between the heart and the aorta [1]. The embryological mechanism responsible for HL is unclear. Lung development begins during the fourth gestational week. HL could result from the non-separation of the splanchnic mesodermal mass [1]. HL usually involves unilateral lung hypoplasia, almost always on the right side [2] (only 10 cases of HL involving left lung hypoplasia have been reported) [2–11]. In this paper, we present another case of HL involving a left hypoplastic lung and perform a systematic review of the previous reports on cases of HL involving left lung hypoplasia.

Case Report

A one-day-old baby boy presented with respiratory problems immediately after birth and was admitted to our neonatal intensive care unit. He was born at 38 weeks and 6 days of gestation by spontaneous transvaginal delivery and had a birth weight of 2,756 g. His Apgar score was 8 at both 1 and 5 minutes. His family history was unremarkable. A ventricular septal defect (VSD), an atrial septal defect (ASD), and pulmonary hypertension were suspected after echocardiography. Chest radiography revealed a hypolucent left lung and mediastinal deviation to the left (Figure 1). Contrast-enhanced computed tomography (CT) was performed with a 128-slice dual-source CT (DSCT) scanner (Somatom Definition Flash, Siemens Healthcare, Forchheim, Germany) and a prospectively echocardiography (ECG)-triggered high-pitch (3.4) spiral acquisition sequence (FLASH Spiral Cardio, Siemens Healthcare, Forchheim, Germany) [12]. The technical parameters were as follows: detector collimation, 128×0.6 mm; section thickness, 0.75 mm; tube current, 146 mAs; and tube voltage, 70 kVp. A 300-mgI/mL non-ionized contrast medium (iohexol: Omnipaque300, Daiichi-Sankyo, Tokyo, Japan) was used. The contrast medium was diluted with saline to 70% (210 mgI/mL). The contrast medium was injected through the right dorsum manus vein at a volume of

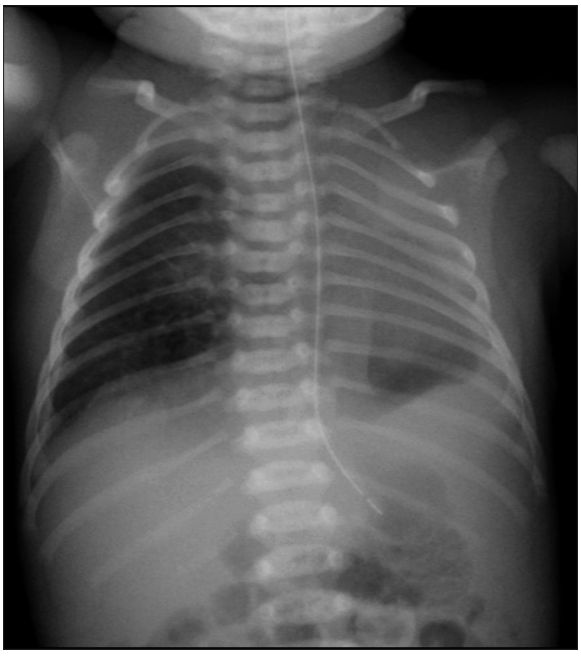

Figure 1. Portable anteroposterior chest radiography revealed a loss of permeability in the left lung field and mediastinal deviation to the left.

2 mL/kg body weight. A saline chaser was not used. The flow rate was fixed at 0.5 mL/s. Bolus tracking was used with scanning by applying the region of interest within the descending aorta at the level of the carina. After the CT number reached 150 HU, a 2-s post-threshold delay was set before the scan. The scan was performed from the supraclavicular to the lower end of the lung without breath holding [13]. The volume CT dose index and dose length product of that scan were 0.42 mGy and 6 mGy·cm, respectively. Sinogram-affirmed iterative reconstruction was performed with a strength value of 3. Multi-planar reconstruction (MPR), minimum and maximum intensity projection (MinIP and MIP), and volume rendering (VR) were used to evaluate the patient's condition.

DSCT revealed left lung hypoplasia and lung parenchymal tissue behind the heart and anterior to the esophagus, and mediastinal deviation to the left (Figure 2). Contrast-enhanced CT (CE-CT) images showed that the left pulmonary artery curved to the right at its origin (Figure 3).

Although left lung agenesis with the protrusion of the right lung into the left thorax was also considered as a differential diagnosis, we diagnosed the patient with HL involving left lung hypoplasia because the left pulmonary artery/vein and left bronchus were confirmed to be present on CE-CT [14]. MinIP and MIP images (Figures 2C, 3B) revealed that the bronchi and pulmonary artery that fed the isthmus of lung parenchymal tissue originated from the left main bronchus and left pulmonary artery, respectively. A VSD and an ASD were also detected on CE-CT with sagittal and axial MPR images (Figure 4). Since the patient's respiratory status remained stable until he was 1 month of age, he was discharged and is currently being followed up as an outpatient.

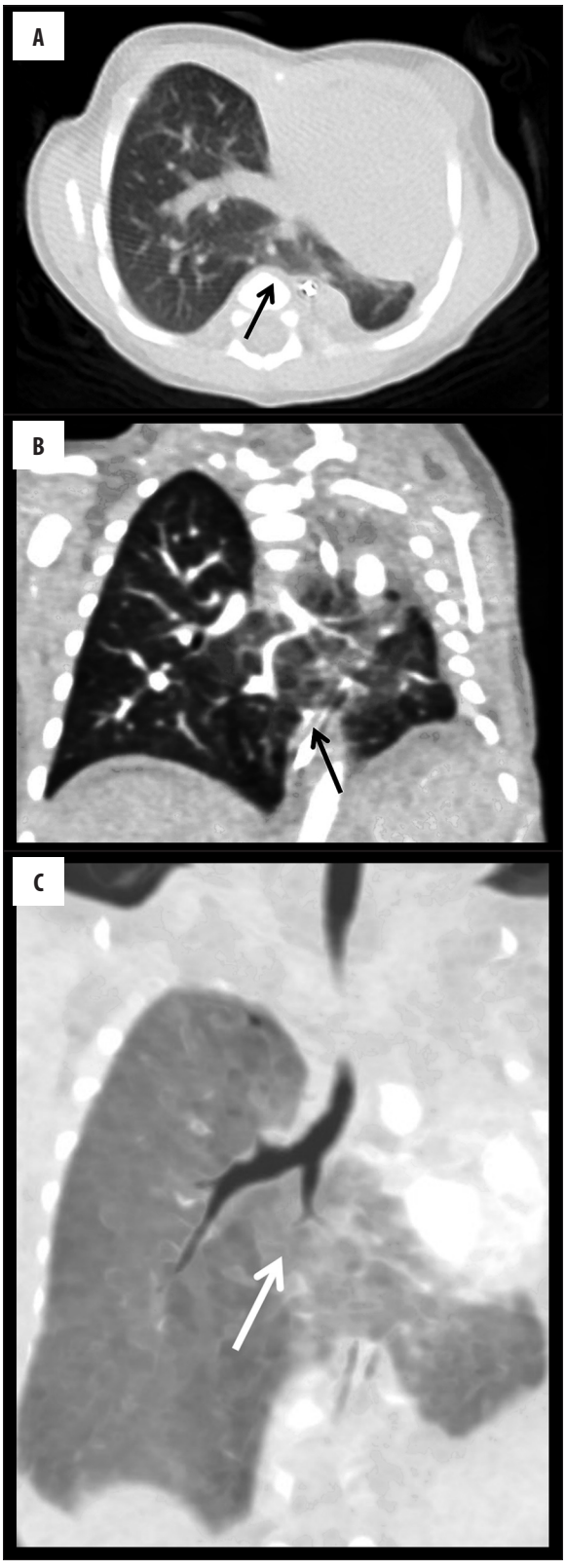

Figure 2. A transverse CT image (**A**) and an MPR coronal image (**B**) depicted the fusion of the posterior parts of the right and left lungs around the ventral esophagus (black arrows). A coronal MinIP image (**C**) demonstrating that the left hypoplastic bronchus had deviated towards the isthmus (white arrow).

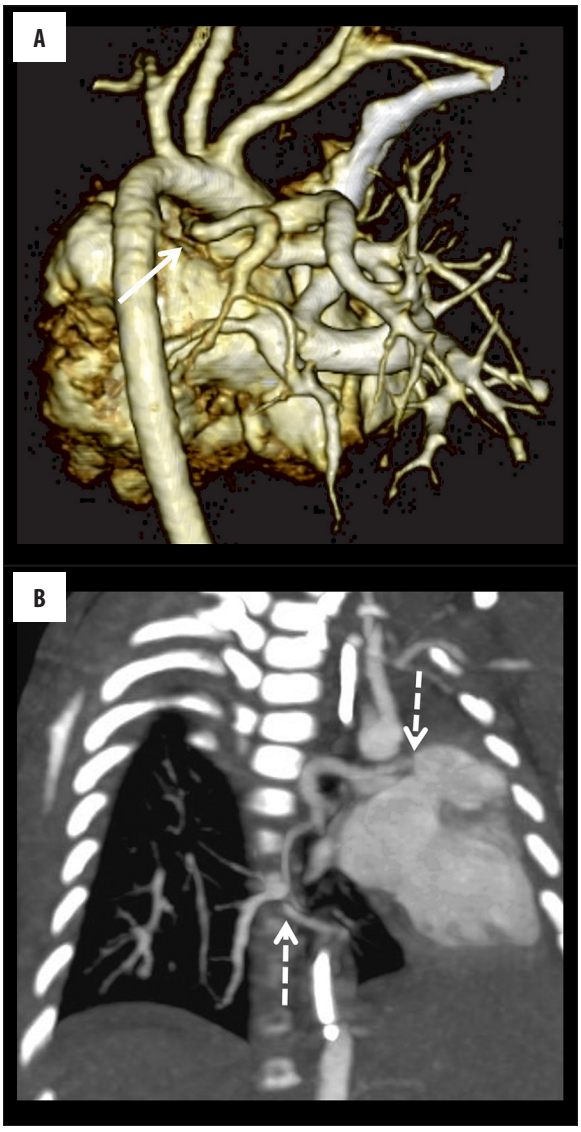

Figure 3. (**A**) The left pulmonary artery was visualized on volume rendering images. The left pulmonary artery curved to the right immediately after it diverged from the pulmonary artery (arrow). (**B**) Coronal MIP images revealed that the pulmonary artery that supplied the isthmus of lung parenchymal tissue originated from the left pulmonary artery (dashed arrows).

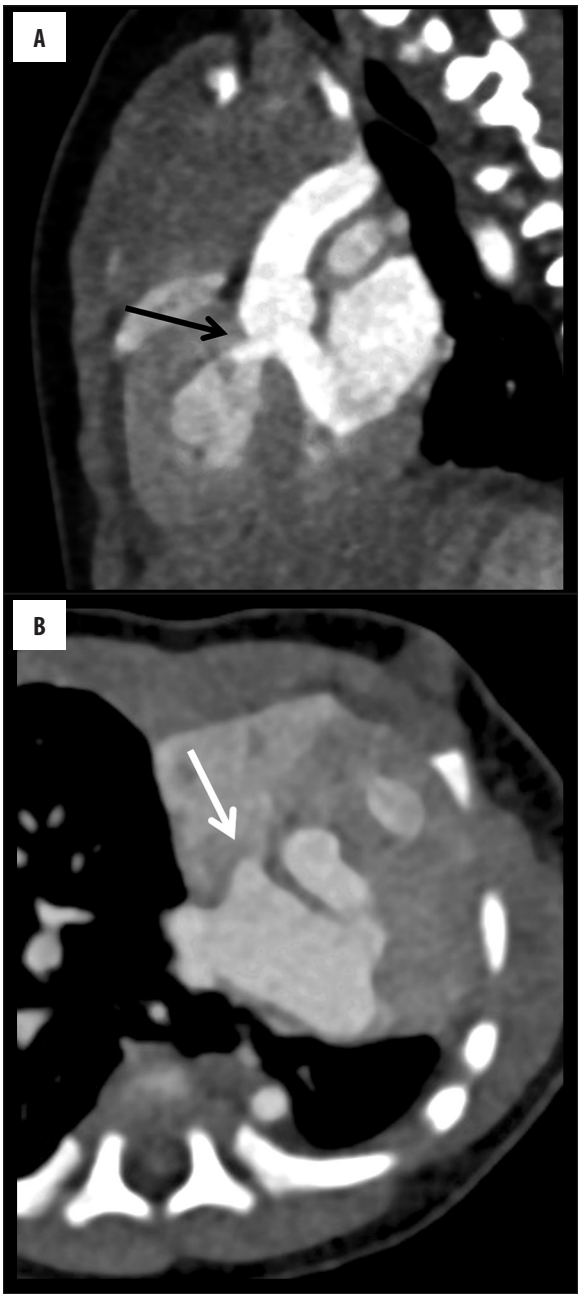

Figure 4. Contrast-enhanced sagittal (**A**) and transverse MRP images (**B**) revealed a VSD (black arrow) and an ASD (white arrow), respectively.

Review of the Literature

We sought to identify all original papers reporting the cases of HL with left pulmonary hypoplasia. Therefore, a comprehensive computer-aided search for relevant primary papers was performed using MEDLINE and PubMed from 1974 to Jun 2015. The following query was used: (horseshoe lung) or (pulmonary hypoplasia). Then, titles and abstracts of the articles were reviewed independently by two researchers (Y.B. and M.N.). Subsequently, full copies of selected articles were retrieved and reviewed by the same two researchers. Only English original papers about human cases of HL with left pulmonary hypoplasia were included. The reports about acquired lesions such as postoperative, postinfectious and posttraumatic changes were excluded. Ten cases of HL involving left lung hypoplasia were found. The fetal autopsy case with HL with left pulmonary hypoplasia reported by Hawass et al. [15] was excluded from this study. Information on those 10 patients and our case are summarized in Table 1 which includes sex, ages, cardiovascular complications, prognosis and used imaging modalities. Abnormal numbers of left pulmonary veins were observed in 2 patients, and cardiovascular malformations such as ASD, VSD and double outlet right ventricle (DORV) were detected in 7 patients. The left pulmonary artery could not be detected in 5 cases [2,4,5,9,10]. Seven cases involved pulmonary hypertension, whereas pulmonary hypertension was not mentioned in the reports

Table 1. Cases of horseshoe lung involving left lung hypoplasia.

First author	Sex	Age	Cardiovascular complications	PH	Prognosis	Imaging modalities
Ersöz A [2]	F	2.5 y	VSD, PFO	(+)	Died	Angiography, Bronchoscopy
Goo HW [3]	M	1 m	ASD, VSD	?	Favorable	Angiography, Bronchoscopy, MDCT (? row), US
Lutterman J [4]	?	3 d	Pulmonary vein stenosis	(+)	Died	Angiography, CT, US
D'Alessandro L [5]	F	4 m	DORV, small left pulmonary vein	?	?	Angiography, Bronchoscopy, CT, MRI, US
Teksam O [6]	F	2 d	ASD, PDA, TAPVC	(+)	?	CT, US
Ogus B [7]	M	8 m	Left pulmonary artery sling	?	?	MDCT (16 row), US
Salerno T [8]	F	10 y	Three left pulmonary veins	(+)	Favorable	Angiography, CT, MRI, US
Jeewa A [9]	M	7 d	HLHS, ASD, VSD	?	Died	MDCT (16 row)
Neves JR [10]	F	3 m	ASD	(+)	Died	Angiography, CT, US
Yildiz AE [11]	M	2 y	Single left pulmonary vein	(+)	?	MDCT (16 row), US
Our case	M	2 d	ASD, VSD	(+)	Favorable	MDCT (128 row), US

M – male; F – female; y – years; d – days, m – months; PH – pulmonary hypertension; PFO – patent foramen ovale; DORV – double outlet right ventricle; HLHS – hypoplastic left heart syndrome; ? – not described; CT – computed tomography (the detail was not described); MDCT – multi-detector row CT, US – ultrasonography, MRI – magnetic resonance imaging.

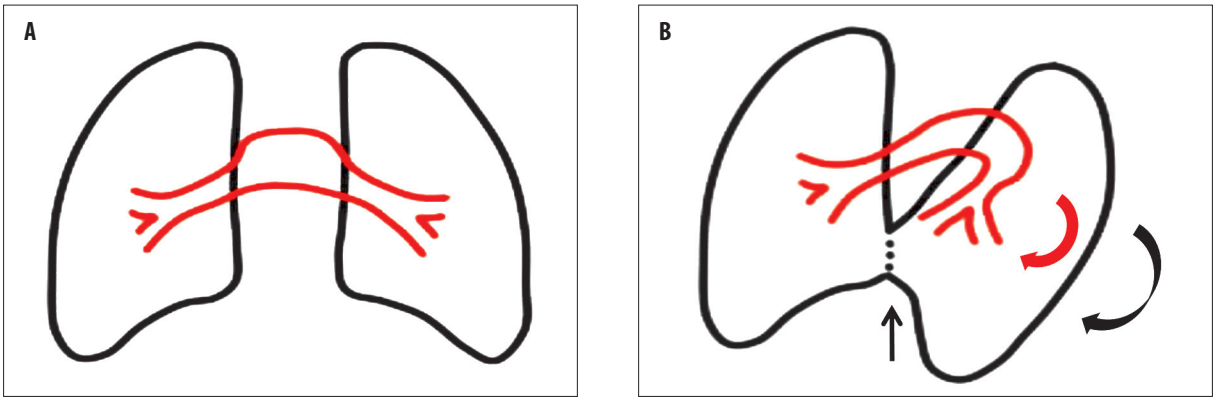

Figure 5. Schema of the lung and pulmonary artery in patients with normal (**A**) and horseshoe lungs (**B**). We postulate that the hypoplastic lung and pulmonary artery might curve and deviate clockwise due to the fusion of the pulmonary parenchyma (curved arrows). Therefore, the pulmonary artery feeding the isthmus of lung parenchymal tissue would arise from the pulmonary artery of the hypoplastic lung (arrow).

on the other 4 cases. Four patients died and 3 survived. In the remaining 4 cases, the patient's prognosis was not reported.

Discussion

The term HL was first used by Spencer [16] in 1962 to describe a malformation characterized by the fusion of the right and left lungs behind the pericardium. The embryological mechanism responsible for HL is unclear. Lung development begins during the fourth gestational week. HL could result from the non-separation of the splanchnic mesodermal mass [2,11].

Mediastinal lung herniation is one of the differential diagnoses of HL. In our case, chest radiography revealed a hypolucent left lung and mediastinal deviation to the left (Figure 1). Mediastinal lung herniation associated with agenesis of the left lung was considered as a differential diagnosis [17]. The pathology of unilateral pulmonary agenesis and hypoplasia has been categorized by Schneider and Schwalbe [14]. In pulmonary agenesis, the lung and bronchi are completely absent on the affected side. On the other hand, lobar agenesis and a hypoplastic lung are seen in lung hypoplasia, but the bronchi and vessels are present [14]. Our patient was diagnosed with HL involving left lung hypoplasia because the patient's left pulmonary artery and vein were clearly identified by 128-slice DSCT (Figure 3). In cases of mediastinal lung herniation, the visceral pleura is located between the herniated and contralateral lungs [17]. In our case, no fissure was detected between the isthmus of lung parenchymal tissue and the left lung on CT images, which was also compatible with HL. In addition, 128-slice DSCT is useful for visualizing the lung parenchyma and determining the patient's cardiovascular configuration. Bronchography and cardiac catheterization used to be necessary for diagnosing HL, but multidetector-row CT (MDCT) has since been found to be useful for this purpose [3].

HL almost always involves unilateral lung hypoplasia and more frequently affects the right side. To the best of our knowledge, only 10 cases of HL involving left lung hypoplasia have been reported [2–11]. In those cases, an abnormal number of left pulmonary veins [8,11] and cardiovascular malformations such as ASD, VSD and DORV were observed [2,3,5,6,9,10]. In our case, an ASD and a VSD were detected with 128-DSCT, but the number of the pulmonary veins was normal. When HL involving left lung hypoplasia is diagnosed, it is important to search for associated cardiovascular malformations.

Among the 10 reported cases in Table 1, the left pulmonary artery could not be detected in 5 cases [2,4,5,9,10]. Jeewa et al. [9] reported that in their case the left pulmonary artery, left pulmonary vein, and left main bronchus were not detected on CE-CT images. Based on the definitions of aplasia and hypoplasia of the lung mentioned above [14], it may be controversial to diagnose those cases as HL involving left lung hypoplasia rather than left lung aplasia. Lutterman et al. [4] and Neves et al. [10] reported that in their case the right pulmonary artery branch ran to the isthmus of the left lung, and the origins of the left pulmonary artery and vein could not be clearly depicted on angiography. Ersöz et al. [2] reported that in their case the left pulmonary artery was not observed on angiography, but small collateral vessels arising from the main pulmonary artery were seen. Among the 10 reported cases of HL involving left lung hypoplasia, the pulmonary arteries of 3 cases [2,4,10] were evaluated with angiography. The absence of the pulmonary artery is considered to be a diagnostic criterion for lung agenesis, but not lung hypoplasia [14]. However, in 4 of the reported cases [2,4,5,10] of HL involving left lung hypoplasia, the left pulmonary artery was not detected. If those 4 cases [2,4,5,10] involved agenesis rather than lung hypoplasia, it is possible that the correct diagnosis would be mediastinal lung herniation associated with left lung aplasia rather than HL. However, we considered that the left pulmonary artery might actually have been present in Ersöz's [2], Lutterman's [4], and Naves's [10] cases. It might have seemed that the left pulmonary artery was absent because it was sought on anterior-posterior projection angiographic images, which could have made branches arising from the left pulmonary artery unclear. In our case, CE-CT revealed that the left pulmonary artery curved to the right at its origin (Figure 3). If we had only examined the left pulmonary artery using anterior-posterior projection angiographic images, then we might have concluded that the left pulmonary artery was absent. We consider that in the cases described by Ersöz [2], Lutterman [4], and Naves [10], the left pulmonary artery might have exhibited a similar shape to that seen in our case. In D'Alessandro's [5] case, the left pulmonary artery was not depicted on CT or MRI, but a small left pulmonary vein and a small left main-stem bronchus were shown. It is uncertain whether that case involved left pulmonary agenesis or left lung hypoplasia. It is more likely that the correct diagnosis was left lung hypoplasia because the left pulmonary vein and main bronchus of D'Alessandro's case were detected [5]. In D'Alessandro's case [5], it is possible that the left pulmonary artery could not be depicted because it was extremely small, and thus, the slice thickness used for CT and MRI might have been too wide.

Regarding the embryological mechanism responsible for kinking of the left pulmonary artery to the right at its origin and then back to the left in peripheral regions, we hypothesized that the pulmonary artery might have curved because the left lung turned clockwise due to pulmonary fusion and hypoplasia (Figure 5). Kinking of the pulmonary artery might tend to be more severe in HL involving left pulmonary hypoplasia than in HL involving right pulmonary hypoplasia because the absence of the right pulmonary artery has not been reported in cases of HL involving right pulmonary hypoplasia. We speculate that this observation is related to the formation of the cardiac loop on the left side on the 24th day of embryo development; due to left pulmonary hypoplasia, the heart might deviate markedly to the left. Lung development begins during the fourth gestational week. HL could result from the non-separation of the splanchnic mesodermal mass [1]. The formation of the pulmonary artery starts from the 6^{th} arch at around the 29^{th} day of embryo development [18]. None of these findings contradict our hypothesis about the development of HL. It is reported that in HL the arterial bronchial supply of the isthmus of lung parenchymal tissue usually originates from the hypoplastic lung [11,19]. The pulmonary artery and bronchus that supply the isthmus of lung parenchymal tissue also originate from the hypoplastic left lung. We consider that this phenomenon can be explained by our hypothesis regarding the deviation of HL (Figure 5).

Cardiovascular malformations, such as ASD and VSD, were reported in each case of HL involving left lung hypoplasia. Pulmonary hypertension occurred in our case and 6 of the previously reported cases [2,4,6,8,10,11] of HL involving left pulmonary hypoplasia. The reports on the other 4 cases [3,5,7,9] did not mention pulmonary hypertension. In 4 of the reported cases of HL involving left lung hypoplasia, the patient died [2,4,9,10]. Therefore, we consider that patients with HL involving left pulmonary hypoplasia are at high risk of pulmonary hypertension. However, as only 10 cases of the condition have been reported, further studies are needed to clarify this issue.

It was reported that 80% of cases of HL involving typical right lung hypoplasia also involved scimitar syndrome, which is a subtype of partial anomalous pulmonary venous connection (PAPVC) in which the abnormal right pulmonary vein drains into the inferior vena cava [19,20]. Scimitar syndrome usually occurs on the right side. However, one left-sided case has been reported [21]. Among the reported cases of HL involving left lung hypoplasia, none of them was associated with scimitar syndrome [2–11], and no scimitar veins were observed in our case.

Dupuis [19] summarized 36 cases of HL, which consisted of 6 cases of HL involving right lung hypoplasia and scimitar syndrome and 30 cases of HL involving right lung hypoplasia that did not involve scimitar syndrome. In their report, the patient died in 67% (4/6) of the cases involving scimitar syndrome and 43% (13/30) of those that did not involve scimitar syndrome. Among the cases of HL involving left lung hypoplasia in which the patient's prognosis was reported, the mortality rate was 57% (4/7) [2,4,9,10]. The reports of 4 cases of HL involving left lung hypoplasia [5–7,11] only described the imaging findings of the condition; i.e., the patient's

prognosis was not stated in those studies. The prognosis of HL involving left lung hypoplasia is probably similar to that of HL involving right lung hypoplasia without scimitar syndrome. Dupuis et al. [19] reported that the prognosis of HL depended on the presence/absence of other malformations and pulmonary hypertension. Therefore, the prognosis of HL might not be affected by whether the hypoplastic lung is right- or left-sided. The accumulation of further cases is needed to shed light on the characteristics of HL.

When HL is diagnosed, it is important to search for complications such as cardiovascular malformations because it has a significant impact on the prognosis of the condition. Symptomatic treatment is usually chosen for pulmonary fusion via the isthmus of lung parenchymal tissue. The indications for surgery include recurrent infections, severe left-to-right shunts (a pulmonary (Qp)/systemic (Qs) blood flow ratio (Qp/Qs) of >2:1,) in the presence of progressive pulmonary hypertension, and cardiovascular malformations [10]. MDCT is useful for detecting cardiovascular malformations such as anomalies of the pulmonary vein or PAPVC [3,20] and for aiding decision-making regarding the optimal treatment strategy for HL. In our case, ASD and VSD were depicted using 128-slice DSCT and a high-pitch spiral acquisition sequence (Figure 4). Nie et al. [12] reported that the coronary artery of children can be depicted using 128-slice DSCT and a high-pitch spiral acquisition sequence. The use of DSCT together with prospective ECG-gating and a high-pitch spiral acquisition sequence is useful for reducing cardiac motion artifacts. Therefore, we considered that DSCT would be useful for evaluating our patient's cardiac anomalies. Since previously reported cases of HL involving left lung hypoplasia were evaluated with angiography and bronchoscopy, their morphological features, e.g., the presence/absence of the left pulmonary artery, were uncertain. Due to the development of MDCT, it is hoped that the pathophysiology of HL involving left lung hypoplasia will be revealed as detailed morphological information gradually accumulates.

Conclusions

HL involving left lung hypoplasia is rare and tends to be associated with pulmonary hypertension and exhibits a poor prognosis. There have not been any reported cases of HL involving left lung hypoplasia associated with scimitar syndrome. MDCT is useful not only for diagnosing HL, but also for searching for associated complications such as cardiovascular malformations. The presence of cardiovascular malformations is a critical factor for decision-making regarding the optimal treatment strategy for HL because it has a significant impact on the prognosis of the condition. In addition, the development of MDCT will hopefully aid the elucidation of the pathophysiology of HL.

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