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Intrathoracic Accessory Liver Tissue and Cleidodysplasia in a Newborn: A rare case report and literature review

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Case Report

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Title:

Intrathoracic Accessory Liver Tissue and Cleidodysplasia in a Newborn:A rare case report and literature review

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Abstract

Intrathoracic accessory liver tissue (IALT) is a rare congenital anomaly that can cause life-threatening respiratory compromise in neonates due to mass effect, yet its association with other skeletal dysplasias remains unexplored. We report the first known case of a neonate presenting with severe respiratory failure caused by a large IALT, concurrently diagnosed with congenital right-sided cleidodysplasia. Preoperative three-dimensional (3D) computed tomography (CT) reconstruction was pivotal in visualizing the vascular pedicle connecting the ectopic tissue to the main liver, effectively differentiating IALT from congenital diaphragmatic hernia (CDH), congenital pulmonary airway malformation (CPAM), and hepatic-pulmonary fusion. Importantly, we propose a novel volumetric ratio—comparing the IALT tissue volume to the right thoracic cavity capacity—to quantitatively assess its impact on respiratory function.

Following stabilization with invasive mechanical ventilation, the patient underwent successful video-assisted thoracoscopic surgery (VATS), which required meticulous dissection of the feeding artery and draining vein while navigating the technical challenges of one-lung ventilation in a neonate. Postoperative histopathology confirmed Type II IALT, and whole-exome sequencing (WES) identified a heterozygous variant in the UGT1A1 gene; however, the pathogenic link between this metabolic gene, IALT, and cleidodysplasia remains unelucidated, suggesting a potential novel syndromic association. This case highlights the critical diagnostic value of 3D CT reconstruction in distinguishing IALT from other thoracic masses and demonstrates the feasibility of VATS for neonatal resection despite technical complexities. The unprecedented co-occurrence of IALT and cleidodysplasia expands the phenotypic spectrum of congenital anomalies, warranting further genetic investigation and rigorous long-term follow-up given the theoretical risk of malignant transformation observed in other ectopic liver tissues. Furthermore, we reviewed and synthesized current literature on pediatric IALT to further clarify its clinical characteristics and the current status of diagnosis and treatment.

Keywords: Intrathoracic accessory liver tissue; Cleidodysplasia; Neonatal respiratory failure; three-dimensional computed tomography reconstruction; Video-assisted thoracoscopic surgery; UGT1A1 variant; Case report.

Background

Intrathoracic accessory liver tissue (IALT) in neonates is rare, and its co-occurrence with cleidodysplasia has not been reported previously. Our center reports the first case. A one-day-old male patient was transferred to our hospital for neonatal respiratory distress. The preoperative diagnosis was congenital diaphragmatic hernia (CDH) with right-sided cleidodysplasia. Surgery was successful, and during the operation, massive accessory liver tissue was found in the right thoracic cavity. Based on imaging studies and postoperative pathological examination, the diagnosis of IALT was made. Postoperative follow-up was conducted for one year, and no recurrence was observed. This case report documents the detailed diagnostic and therapeutic

process, which can serve as a reference for the diagnosis and treatment of IALT in neonates, reducing the risk of misdiagnosis.

Case report

The patient was a male term infant, born via spontaneous vaginal delivery at 39 weeks of gestation to a gravida 1, para 1 (G1P1) mother, with a birth weight of 2.6 kg. He was transferred to Kunming Children's Hospital on day one of life due to severe respiratory distress. Immediately after birth at the referring hospital, the infant exhibited cyanosis, tachypnea, and expiratory grunting; however, no convulsions or high-pitched crying were observed. Although neonatal respiratory distress syndrome (NRDS) was initially suspected, a chest computed tomography (CT) scan revealed findings suggestive of CDH, prompting immediate transfer to our institution. Upon arrival, invasive mechanical ventilation was initiated. Physical examination revealed a critically ill infant with poor responsiveness, mild icterus, and central cyanosis. Significant subcostal, intercostal, and suprasternal retractions were noted during inspiration. Musculoskeletal examination showed a prominent right acromion. Head examination revealed a normal head shape with a patent anterior fontanelle measuring approximately 2.5 cm × 2.5 cm and normal tension. Auscultation of the lungs revealed no rales. Cardiac examination demonstrated a regular rhythm with strong heart sounds and a grade II/VI systolic murmur heard over the precordium. The abdomen was soft and non-distended, with no palpable hepatosplenomegaly and normal bowel sounds. Extremities were warm without mottling; however, the capillary refill time (CRT) at the heel was prolonged at 3 seconds. Prior to transfer, the infant had received continuous positive airway pressure (CPAP) and dopamine for inotropic support.

On admission, laboratory investigations yielded the following results: Complete blood count (CBC) revealed a white blood cell (WBC) count of 14.33×10^9/L (neutrophils: 8.46×10^9/L), red blood cell (RBC) count of 4.85×10^{12}/L, and platelet count of 372×10^9/L. The C-reactive protein (CRP) level was < 0.2 mg/L. Liver function tests (LFTs) showed an alanine aminotransferase (ALT) level of 6 U/L, aspartate aminotransferase (AST) of 28 U/L, total bilirubin of 178.8 μmol/L (direct: 5.4 μmol/L; indirect: 173.4 μmol/L), and albumin of 32.5 g/L. Imaging studies were pivotal for diagnosis. CT of chest identified a solid mass measuring 31 × 33 × 42 mm within the right thoracic cavity. The IALT demonstrates enhancement patterns synchronous with the main liver parenchyma across all phases (**Fig. 1A–D**).Notably, concurrent chest radiography confirmed the absence of the right clavicle (**Fig. 2**). A contrast-enhanced gastrointestinal study was unremarkable.

The significant mass effect exerted by the large space-occupying lesion in the right thoracic cavity precipitated severe neonatal respiratory distress, necessitating immediate invasive mechanical ventilation. Following preoperative optimization and stabilization of respiratory and

hemodynamic parameters, Video-assisted thoracoscopic surgery (VATS) was performed upon obtaining parental informed consent. The procedure was conducted under single-lung ventilation with selective left-sided intubation. Intraoperatively, all three lobes of the right lung were observed to be compressed superiorly by the intrathoracic mass (**Fig. 3A**). A firm, brownish, solid mass measuring 30 × 40 × 45 mm was visualized on the diaphragmatic surface . Its vascular supply originated directly from the diaphragm; notably, no diaphragmatic defect was identified (**Fig. 3B**). The feeding arteries and veins were meticulously dissected, ligated, and divided (**Fig. 3C**), allowing for complete excision of the mass, which was subsequently sent for histopathological examination (**Fig. 4**). Prior to chest closure, the anesthesiologist performed a lung recruitment maneuver, resulting in satisfactory re-expansion of the right lung.

Histopathological examination revealed hepatic lobular architecture with intervening portal tracts. Microscopic findings included focal hepatocellular edema, spotty necrosis, and small foci of extramedullary hematopoiesis. Additionally, a mixed acute and chronic inflammatory infiltrate was observed. Scattered hepatocytes exhibited nuclear atypia, characterized by enlargement, irregularity, and hyperchromasia. Immunohistochemical analysis demonstrated the following profile: Cytokeratin 8/18 (CK8/18) positive; Cytokeratin 19 (CK19) and Cytokeratin 7 (CK7) positive in bile ducts; and Cluster of Differentiation 68 (CD68) positive in scattered cells (**Fig. 5A–D**). Stains for Human Melanoma Black 45 (HMB-45), Melanoma Antigen Recognized by T cells 1 (Melan-A), Thyroid Transcription Factor 1 (TTF-1), and Cytomegalovirus (CMV) were negative.

Invasive mechanical ventilation was discontinued on postoperative day 1. Follow-up liver function tests performed two weeks postoperatively showed normalization of hepatic enzymes [ALT: 13 U/L; AST: 24 U/L] and albumin (41.2 g/L). Total bilirubin was 62.2 μmol/L, predominantly indirect (57.2 μmol/L), with a direct fraction of 5.0 μmol/L. The patient was discharged in stable condition. Although readmitted once at three months post-discharge for pneumonia, the patient remains asymptomatic and well at the one-year follow-up.

Discussion

Ectopic liver tissue (ELT) Based on anatomical connectivity, ELT is traditionally categorized into two forms: an accessory liver lobe, which maintains vascular or biliary continuity with the main liver, and ectopic liver proper, defined as a distinct nodule completely separated from the main organ and supplied by local aberrant vessels [1]. Although the exact pathogenesis remains elusive, prevailing hypotheses suggest it arises from abnormal migration of hepatic progenitors during embryogenesis or the displacement of hepatic cell clusters into the thoracic cavity during diaphragmatic closure [2, 3].

While ELT most frequently occurs within the abdominal cavity (typically adjacent to the gallbladder, spleen, or adrenal glands), IALT is a rare entity. IALT is often discovered incidentally on imaging and has been documented primarily in isolated case reports [3–8]. To standardize the characterization of IALT, Adin et al. [7] proposed a three-tiered classification system: Type I, where intrathoracic tissue connects directly to the main liver through a diaphragmatic defect; Type II, characterized by a connection via a vascular pedicle; and Type III, representing a completely isolated nodule with an independent vascular supply and drainage. Herein, we report a case of Type II IALT, the most prevalent subtype of this rare condition.

Pediatric cases of IALT are exceptionally rare. A comprehensive literature review identified only 13 reported cases of pediatric IALT (**Table 1**)[9,12-22]. Stratified by Adin's classification, six cases were Type II, six were Type III, and one lacked subtype specification [6,7,10,11]. Anatomically, the lesions predominated in the left thoracic cavity (9/13) compared to the right (4/13). Notably, seven of these cases occurred in the neonatal period, where respiratory distress emerged as the predominant clinical manifestation. While IALT is frequently described as an asymptomatic incidental finding in the general population, our review highlights a distinct age-dependent symptomatic profile. All neonates in the reviewed cohort required preoperative mechanical ventilation, likely attributable to the limited thoracic compliance in this age group combined with the mass effect of the ectopic tissue compressing adjacent lung parenchyma. In contrast, the six non-neonatal cases presented with milder respiratory symptoms, such as cough and pneumonia.

Preoperatively, IALT is often misdiagnosed as CDH, with congenital pulmonary airway malformation (CPAM) and pulmonary sequestration serving as key differential diagnoses. Surgical resection was performed in all reported cases, with one recorded mortality. Intraoperatively, coexisting CDH was confirmed in 8 of 13 patients (61.5%); additional associations included intralobar pulmonary sequestration and pulmonary mesenchymal hamartoma .

The clinical impact of IALT appears to be governed by the ratio of lesion volume to hemithoracic capacity rather than absolute size. Due to the restricted neonatal thoracic volume, a lesion of a given size may precipitate severe respiratory distress requiring ventilatory support in a neonate, whereas the same lesion might manifest only as a mild cough in a toddler [13, 14]. Consequently, we propose that the IALT-to-hemithorax volume ratio serves as a more accurate predictor of respiratory compromise. In our present case, the IALT occupied 42.48% of the right hemithorax, correlating with the patient's presentation of significant respiratory distress.

We present a case of neonatal IALT located in the right hemithorax. The infant exhibited progressively worsening respiratory distress immediately after birth. 3D CT reconstruction of the chest reconstruction revealed a well-circumscribed, round mass measuring 4.0 cm in diameter.

Crucially, the 3D visualization delineated a vascular pedicle traversing the diaphragm, connecting the ectopic tissue to the main liver; the feeding artery originated from the abdominal aorta, while venous drainage communicated with the portal venous system (**Fig. 6A–D**). This detailed vascular mapping not only facilitates a definitive diagnosis but also provides essential guidance for surgical planning.

The substantial volume of the IALT exerted a significant mass effect within the confined neonatal thoracic cavity, directly compromising pulmonary expansion and resulting in severe respiratory distress. While the pathophysiological mechanism of compressive lung hypoplasia mirrors that of CDH, the anatomical origins remain distinct [23]. These findings underscore the importance of including IALT in the differential diagnosis of intrathoracic masses identified during prenatal ultrasound screening. Specifically, the observation of a thoracic mass maintaining continuity with the diaphragm or abdominal structures should heighten clinical suspicion for this rare entity, prompting further vascular imaging to confirm the diagnosis.

Due to its significant mass effect, large IALT in neonates can directly compress the lung parenchyma and mediastinum, precipitating progressive NRDS, necessitating immediate mechanical ventilation. Clinical manifestations are invariably more severe in neonates compared to older children or adults, attributable not only to the restricted neonatal thoracic volume but also to the potential for rapid lesion growth. For instance, a previously reported case described a large intrathoracic mass discovered six years following neonatal repair of CDH, which was subsequently confirmed as Type II IALT upon resection [12]. Furthermore, imaging findings of a large IALT mass with a narrow vascular pedicle raise concerns for pedicle torsion, a catastrophic complication that warrants heightened vigilance [24]. Consequently, symptomatic neonatal IALT should be regarded as a surgical emergency. We recommend definitive resection once respiratory and hemodynamic stability is achieved to prevent irreversible pulmonary hypoplasia.

Regarding long-term outcomes, ectopic liver tissue located within the abdominal cavity is known to carry a risk of malignant transformation. Liu et al. [25] documented a case of hepatocellular carcinoma arising in pancreatic ectopic liver tissue, while Akbulut et al. [26], in a review of 91 cases of gallbladder ectopic liver, identified one instance of malignant degeneration. To date, no cases of malignant transformation specifically involving IALT have been reported in the literature. However, given the established oncogenic potential of ectopic hepatic tissue elsewhere, this risk cannot be dismissed solely based on the current absence of reports. The lack of pediatric cases may simply reflect insufficient follow-up duration rather than a true absence of risk. Therefore, lifelong surveillance is prudent. In line with this rationale, our patient will undergo rigorous long-term follow-up to monitor for any late-onset complications.

Preoperative diagnosis of IALT remains a significant clinical challenge, often eluding definitive confirmation until postoperative histopathological examination. The radiological presentation of IALT frequently overlaps with more common congenital anomalies, necessitating a careful differential diagnosis that includes CDH and CPAM. Furthermore, distinction must be made from the rare entity of hepatic-pulmonary fusion, which can mimic IALT with strikingly similar clinical and imaging features [27, 28]. While advanced imaging techniques such as 3D CT reconstruction can strongly suggest the diagnosis by visualizing the characteristic vascular pedicle, histological verification remains the gold standard for final confirmation.

Notably, this patient presented with a concurrent anomaly: congenital right-sided cleidodysplasia, evidenced by radiographic discontinuity of the right clavicle. While isolated clavicular defects are a hallmark of cleidocranial dysplasia (CCD) (an autosomal dominant disorder caused by RUNX2 variants affecting cranial, clavicular, and dental development)[29], our patient lacked the other characteristic features of CCD. From an embryological perspective, a direct link between these anomalies seems unlikely: the liver originates from foregut endoderm, whereas the clavicle develops via intramembranous ossification of mesodermal mesenchymal cells [30]. A comprehensive literature search confirmed no prior reports of IALT coexisting with cleidodysplasia, suggesting a potentially novel syndromic association.

To explore a potential genetic etiology, whole-exome sequencing (WES) was performed following ethical approval (Kunming Children's Hospital, Approval No. 2026–05-065-K01) and informed parental consent. WES identified a heterozygous variant in the UGT1A1 gene, which encodes bilirubin UDP-glucuronosyltransferase, a key enzyme in hepatic bilirubin metabolism [31, 32]. While UGT1A1 deficiencies are well-established causes of Gilbert syndrome and neonatal hyperbilirubinemia, no known association exists between this gene and skeletal development or hepatic ectopia. Consequently, the pathogenicity of this variant in the context of our patient's multisystem anomalies remains unelucidated. Nevertheless, the unprecedented co-occurrence of IALT and cleidodysplasia challenges current understanding of developmental biology. This case highlights the complexity of multisystem malformations and suggests that even genes with established metabolic functions may harbor undiscovered pleiotropic effects or serve as markers for broader genomic instability. As the first reported instance of this constellation of findings, it offers a novel perspective for future research into the genetic underpinnings of rare developmental anomalies.

Conclusion

This report documents the first known case of IALT coexisting with congenital cleidodysplasia, a novel association that warrants further genetic investigation. Clinically, neonatal IALT typically presents with severe respiratory failure due to mass effect, necessitating prompt invasive mechanical ventilation. We emphasize that preoperative 3D CT reconstruction of the chest is indispensable for diagnosis. It clearly delineates the vascular pedicle connecting the ectopic tissue to the main liver, which is a key feature distinguishing IALT from CDH and

hepatic-pulmonary fusion. While VATS offers a minimally invasive approach, it presents significant technical challenges in neonates, particularly regarding one-lung ventilation; thus, surgical resection should be pursued only after stabilizing respiratory and circulatory function. Meticulous dissection of the feeding artery and draining vein is critical to prevent hemorrhage, and definitive diagnosis remains dependent on postoperative histopathology. Although malignant transformation of IALT has not yet been reported, the potential oncogenic risk observed in other ectopic liver tissues underscores the necessity for rigorous long-term follow-up. Ultimately, this unique case expands the phenotypic spectrum of IALT and highlights the importance of considering multisystem anomalies in rare congenital disorders.

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Declarations

Acknowledgment

Not applicable.

Authors' contributions

MXZ, GZY and WH designed the study; MXZ, SQP, XXW, KC, WHZ, JYF collected and analyzed the data; MXZ and TY drafted the initial manuscript; MXZ, TY, ZML revised the article critically; MXZ, WH and WH reviewed and edited the article; MXZ are first authors. GZY and WH are co-corresponding author. All authors approved the final manuscript.

Ethics approval and consent to participate

This study was approved by the ethical committee of Kunming Children's Hospital (2026-05-065-K01); This study is by the relevant guidelines and regulations. The data in this study were obtained from this patient and his legal guardian. Written informed consent was obtained from the patient's parents. Informed consent was obtained from the patient's parents and/or legal guardians for the publication of identifying information/images in online open access publications.

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Consent for publication

Informed consent was obtained from the patient's parents and/or legal guardians for the publication of identifying information/images in online open access publications.

Availability of data and materials

Data will be available from the corresponding author on a reasonable request.

Competing interests

The authors of this manuscript declare no conflict of interest.

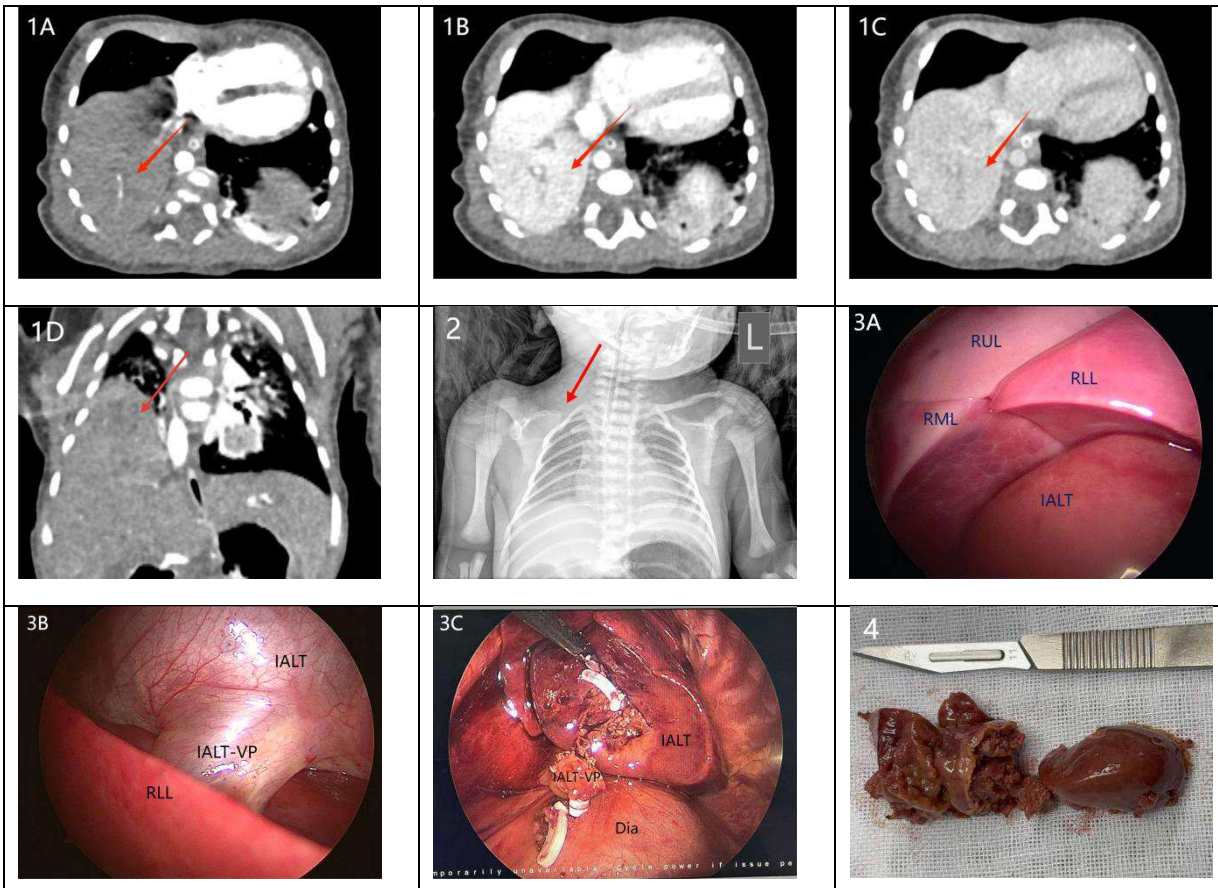


Fig.1 Preoperative imaging findings: **1A-1C** Contrast-enhanced computed tomography of the abdomen and chest in the arterial (**1A**), portal venous (**1B**), and delayed (**1C**) phases. **1D** Sagittal reconstruction of the chest CT clearly delineates the IALT located in the right hemithorax. **Red arrows** indicate the IALT mass. **Fig.2** Frontal chest X-ray reveals discontinuity and hypoplasia of the right clavicle, The **red arrow** points to the affected segment of the right clavicle. **Fig.3** Intraoperative findings during VATS: **3A** Visualization of the IALT situated in the right costophrenic angle, inferior to the right lung lobe and superior to the intact diaphragm. **3B** Exposure of the vascular pedicle (VP) connecting the IALT to the abdominal liver. The pedicle is observed traversing the diaphragmatic hiatus, confirming the transdiaphragmatic blood supply. **3C** Completion of the resection following ligation of the feeding vessels with a biological hemostatic clip and transection of the vascular pedicle. The IALT is completely separated from the surrounding structures. **Fig.4** The isolated IALT specimen following resection.

IALT: intrathoracic accessory liver tissue; VATS ： video-assisted thoracoscopic surgery; RUL: right upper lobe; RML: right middle lobe; RLL: right lower lobe; IALT-VP: the vascular pedicle of the IALT; Dia: diaphragm.

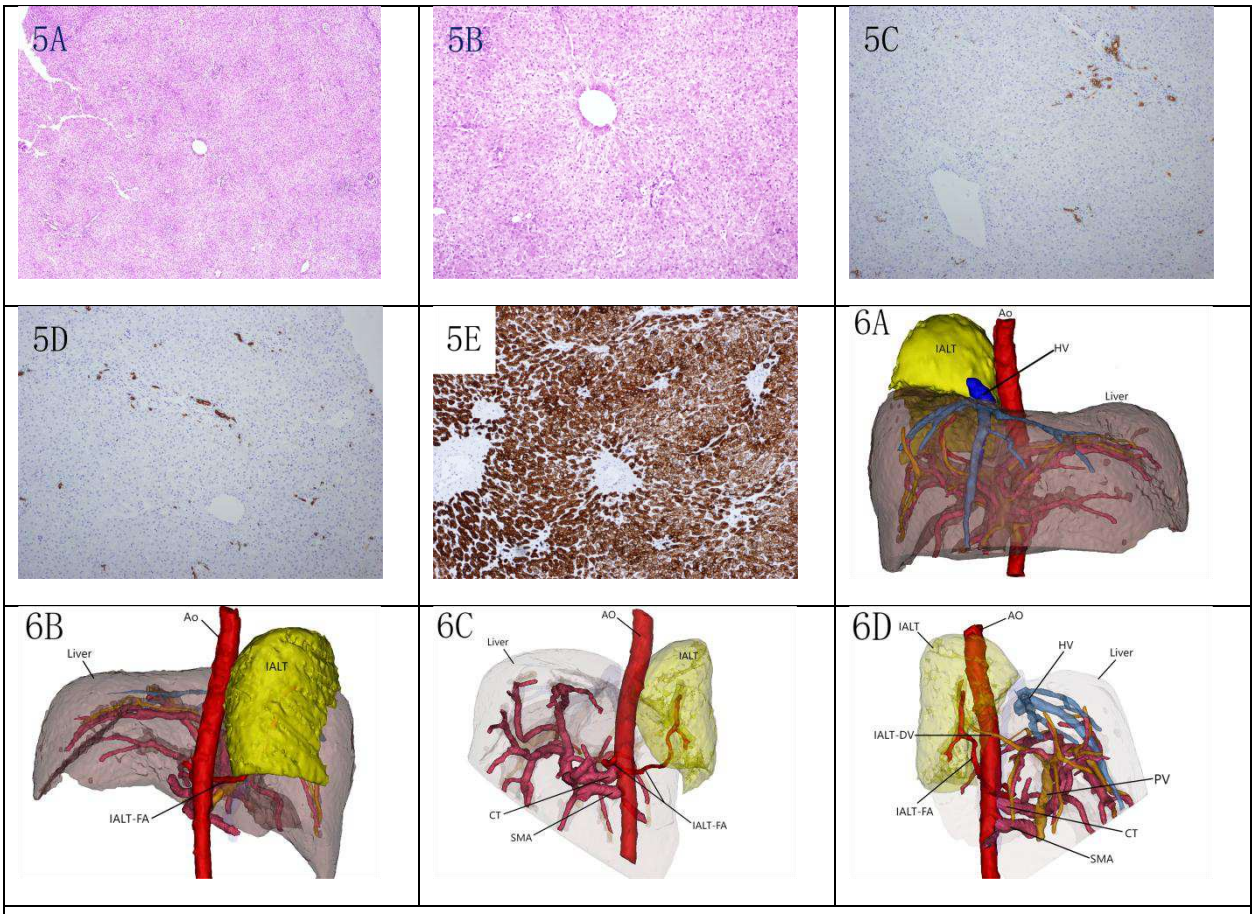


Fig.5 Postoperative pathological examination and immunohistochemical results: **5A** Preserved hepatic lobular architecture, with hepatocytes arranged in cords radiating from the central veins (HE, 40X). **5B** Portal tracts are observed in the periphery (HE, 100X). **5C-5E** Postoperative immunohistochemical findings (**C:** CK7, **D:** CK19, **E:**CK8/18 ）（C-E: 100X） .**Fig.6** Three-dimensional reconstruction of chest computed tomography Imaging findings. **6A-6B** The IALT located within the right hemithorax, situated superior to the diaphragm. The diaphragmatic contour remains intact. **6C** Arterial phase reconstruction identifies the feeding artery of the IALT originating directly from the supraceliac abdominal aorta, just superior to the celiac trunk. **6D** Venous phase reconstruction delineates the draining vein, which courses inferiorly through the diaphragm to drain into the portal venous system of the main liver.

IALT: intrathoracic accessory liver tissue; Ao: Aorta; HV: hepatic Vein; CT: celiac trunk; SMA: superior mesenteric artery; PV: portal vein; IALT-FA: feeding artery of IALT; IALT-DV: draining vein of IALT.

Table 1 Literature review of published cases associated with IALT in neonates

References	Sex/ age	Ty- pe	Symptoms	Preoperative diagnosis	mechanical ventilation	Location （thoracic cavity）	Clinical management	The size of IALT (cm)
Luoma R et al.(15)	F/1d	II	pleural effusion, respiratory distress	pleural effusion	Yes	Left	Surgery, IALT without CDH	3.0×1.5×0.8
Beiler HA et al.(16)	M/1 d	III	respiratory distress, acidosis	CDH	Yes	Left	Surgery;IALT with CDH	5.5×1.5×1.0
Mito K et al.(17)	M/4 d	III	respiratory distress	CDH	Yes	Left	Surgery ; IALT with CDH	1.3
Mito K et al.(17)	M/4 d	III	respiratory distress	CDH	Yes	Left	Surgery ; IALT with CDH	1.0×0.4×0.3
Shah KD et al.(13)	F/1d	II	respiratory distress	CDH	Yes	Left	Died during the operation, IALT with CDH	5.0×4.0×2.0
Ortiz Otero A et al.(9)	F/1d	N	respiratory distress	CDH	Yes	Left	Surgey,Hepatic mesenchymal hamartoma arising in IALT,CDH	3.5
Antoniou D et al.(18)	F/14 d	III	N/A	CDH	Yes	Left	Surgey,Cystic mesenchymal hamartoma arising in IALT,CDH	2.0×2.5×1.0
Bedii Salman A et al.(19)	N/6 mo	II	N/A	CDH	N/A	Left	Surgery;IALT with CDH	6.0×8.0
Chen F et al.(20)	M/13mo	II	pneumonia	EPS	No	Right	Surgery	6.0×2.5×3.0

Babu R et al.(21)	M/18mo	II	mild respiratory distress	CDH	No	Right	Surgery , IALT without CDH	8.0×5.0×5.0
Iber T et al.(12)	M/6 yr	II	mild asymmetry of the chest	recurrent CDH	No	Right	Surgery, Postnatal surgical repair of CDH	8.0
Choi SU et al.(14)	M/3 yr	III	Cough	EPS and CPAM	No	Right	Surgery;IALT with IPS	4.7×3.5×3.0
ZOU CC et al.(22)	M/4 yr	III	N/A	EPS	No	Left	Surgery	2.2×1.2×0.8
Current Study	M/1 d	II	respiratory distress	CDH	Yes	Right	microsurgery , IALT without CDH	6.0×5.0×4.5

F : Female M : Male ; cm : centimeter ; d : days ; mo : months ; yr : years ; N/A : information not available ; cm : centimeter ; CDH : congenital diaphragmatic hernia ; IALT : intrathoracic accessory liver tissue ; EPS : extralobar pulmonary sequestration ; IPS : Intralobar pulmonary sequestration ; CPAM : congenital pulmonary airway malformation.