

Can Chest CT Differentiate Between Low-Risk and Higher Risk Congenital Lung Lesions in Infants?

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Abstract

Purpose

To determine whether chest CT can differentiate low-risk congenital lung lesions (pulmonary sequestration and CPAM type 2) from higher-risk lesions (CPAM type 1 and pleuropulmonary blastoma [PPB]) and thereby support the selection of patients for conservative management.

Methods

In this retrospective study (2003–2024), we identified infants with congenital lung lesions who underwent chest CT and subsequent pathologic confirmation. Two pediatric radiologists independently reviewed the anonymized CT scans, blinded to clinical and pathologic data. Lesions with a systemic arterial supply or predominant hyperinflation were classified as low risk; those with a solitary cyst, a cyst > 2 cm, or a complex cyst without systemic supply were classified as higher risk. Diagnostic confidence was graded as low, moderate, or high. Pathologic diagnosis served as the reference standard, and Cohen's kappa was used to assess interobserver agreement.

Results

Eighty-three children (52 male) were included. Low-risk lesions (n = 73) comprised CPAM type 2 with or without sequestration (n = 51), isolated sequestration (n = 18), and other benign lesions (n = 4). Higher-risk lesions (n = 10) included CPAM type 1 (n = 8) and type I PPB (n = 2). For moderate-to-high-confidence diagnoses, sensitivity was 100% for both radiologists; specificity was 89.2% and 93.1%; and the negative predictive value was 100%. Interobserver agreement was almost perfect (98.4%; $\kappa = 0.95$).

Conclusion

Chest CT enables reliable differentiation of higher-risk from low-risk congenital lung lesions and can guide the selection of candidates for conservative management.

Introduction

With routine fetal ultrasound (US) screening complemented with fetal MRI (fMRI), congenital lung lesions are more commonly discovered and most of them are asymptomatic creating new management challenges^{1–4}. The most common lung lesions are congenital pulmonary airway malformation (CPAM) and bronchopulmonary sequestration. CPAM encompasses a group of lesions arising from abnormal lung development, and its classification has been revised several times. Initially, CPAM was divided into five types (type 0 to type 4) under the Stocker system^{5,6}. Type 0 is now designated congenital acinar dysplasia, which is incompatible with life, and type 4 has been reclassified as type I pleuropulmonary blastoma (PPB)^{7,8}.

CPAM type 1 results from a somatic mutation in KRAS (Kirsten rat sarcoma viral oncogene homolog), a gene encoding a protein involved in the signaling pathways that regulate cell growth, differentiation, and apoptosis.

Most CPAM type 1 lesions harbor mucinous cell clusters and carry a low risk of malignant transformation to mucinous adenocarcinoma^{7,9}. In addition, small series and case reports on late presentation of CPAM suggest that CPAM type 1 carries a higher risk for superimposed infection^{10–13}. Historically, CPAM type 1 was considered the most common subtype; however, with near-universal prenatal US screening and increased lesion detection, type 2 is now recognized as the most prevalent subtype¹⁴. CPAM type 2 is an acquired lesion resulting from interrupted lung development secondary to bronchial obstruction or atresia and is not associated with malignant transformation^{5,7}. Type 3 is a solid, mass-like lesion with an overgrowth of uniform bronchiolar-like structures and is rare⁷.

Bronchopulmonary sequestration is an isolated mass of lung with no bronchial communication with a systemic blood supply. A sequestration may be enclosed within its own pleura (extralobar sequestration) or lie within the visceral pleura (intralobar sequestration)⁵. Sequestrations are often associated with CPAM type 2, and the combined lesion is termed a hybrid lesion^{5,15}. Both extralobar and intralobar sequestration are also regarded as consequences of bronchial atresia⁷.

Surgery is indicated for symptomatic lung lesions, which most commonly present as respiratory distress in the newborn period or as superimposed infection in older children. However, no consensus exists regarding the management of asymptomatic lesions. Early surgical intervention has been advocated to eliminate infection risk and to prevent the rare possibility of malignant transformation, and resection during infancy may also promote better compensatory lung growth. These considerations must be weighed against the evidence that most CPAMs and sequestrations decrease in size or involute during conservative follow-up^{4,16,17}.

Type I PPB is a malignant lung tumor that most commonly presents in infants (median age, 9 months) and frequently mimics a developmental lung cyst. It is characterized by large cysts (2–16 cm), often multiseptated, with primitive mesenchymal cells beneath benign epithelium that sometimes contain rhabdomyoblasts or cartilage nodules. Type I PPB may be solitary, multiple, or bilateral and carries a favorable prognosis, with 85–90% overall survival. However, it may progress to the more aggressive type II or type III PPB, which are typically diagnosed in older children (median age, 36 months for type II and 42 months for type III). Recognizing type I PPB as a neoplasm with malignant potential is essential for appropriate treatment and follow-up^{2,18,19}.

Although certain imaging features favor CPAM over type I PPB—including single-lobe location, absence of pneumothorax, simple cysts, and hyperinflation^{2,20}, there is some overlap in imaging particularly between CPAM type 1 and PPB, and one study has shown that imaging cannot reliably differentiate these two entities²¹.

Recognizing this imaging overlap between CPAM type 1 and type I PPB, we focused on differentiating clinically relevant risk categories rather than individual histologic diagnoses. This distinction is particularly important when conservative management is being considered. CPAM type 1 is associated with an increased risk of superimposed infection and a rare risk of malignant transformation, whereas type I PPB is a neoplasm that warrants surgical resection. In contrast, CPAM type 2 and pulmonary sequestration carry a lower risk of infection and no known risk of malignant transformation.

Methods

We retrospectively evaluated a cohort of patients over a 21-year period (2003–2024) to determine whether chest CT imaging characteristics can reliably distinguish low-risk lesions (sequestration and CPAM type 2) from higher-risk lesions (CPAM type 1 and PPB). From the radiology and pathology information systems, we retrieved all children who underwent surgery for a congenital lung lesion and had a postnatal chest CT within the first 12 months of life. From the electronic medical record, we retrieved demographic data, gestational age at birth, newborn respiratory symptoms, and associated medical or chromosomal abnormalities.

Review by radiologists

Each radiologist evaluated lesion location and imaging characteristics, including mediastinal shift, atretic bronchus, systemic feeding artery, cystic components (solitary or multiple [> 4]), and diaphragmatic hernia. When a cystic lesion was identified, it was further characterized as simple or complex, with notation of septations, a dominant cyst (defined as a single large cyst surrounded by smaller cysts), and solid components.

Each radiologist assigned an imaging diagnosis and an overall level of diagnostic confidence (low, moderate, or high) for each case. Imaging diagnoses were categorized as follows. A higher-risk lesion was defined by the presence of a solitary cyst, a complex cyst without a systemic feeding artery, or a cyst with a maximum diameter > 2 cm. A lower-risk lesion was defined by any cyst with a systemic feeding artery or a lesion with predominant hyperinflation. Discrepancies in risk classification between the two radiologists were resolved by consensus.

Pathology criteria for congenital lung lesions

CPAM type 1 was diagnosed on the basis of cysts lined by undulating, pseudostratified, ciliated columnar epithelium, with or without mucinous cell clusters⁷. CPAM type 2 was diagnosed on the basis of bronchiolar architecture characterized by cuboidal-to-columnar epithelium without mucinous cell clusters⁷. Pulmonary sequestration was diagnosed on the basis of a systemic arterial supply and the absence of communication with the tracheobronchial tree, with or without associated CPAM type 2 (hybrid lesion)²². PPB was diagnosed on the basis of a multilocular cyst lined by low cuboidal-to-attenuated alveolar-type epithelium, containing primitive mesenchymal cells within the cyst walls and well-formed cambium layers¹⁸.

Cases with incomplete histologic characterization in the original pathology reports were reviewed by a pediatric pathologist.

Statistical Analysis

Univariable chi-square analysis was performed separately for each radiologist to estimate risk ratios for higher-risk pathology (CPAM type 1 or PPB) associated with the following imaging features: multilobar versus single-lobe involvement, mediastinal shift, hyperinflation, atretic bronchus, systemic feeding artery, cystic lesion, multiple cysts (> 4), simple cyst morphology, septated cyst morphology, dominant cyst, solid cyst component, maximum cyst diameter, and diaphragmatic hernia. The features that best distinguished low-risk from higher-risk lesions were then identified.

For each radiologist and for the consensus interpretation, the sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of the assigned risk categories (low risk, higher risk) were calculated using pathologic diagnosis as the reference standard. Indeterminate-risk and low-confidence evaluations were excluded from this analysis. A moderate- or high-confidence diagnosis of low risk was considered a true positive when pathology demonstrated pulmonary sequestration, CPAM type 2, or other pathology not associated with malignant transformation, and a false negative when pathology demonstrated CPAM type 1 or PPB. A moderate- or high-confidence diagnosis of higher risk was considered a true positive when pathology demonstrated CPAM type 1 or PPB, and a false positive when pathology was not associated with malignant transformation.

Interobserver agreement between radiologists for both the specific diagnosis and the risk-category assignment was assessed using Cohen's kappa, with agreement categorized as follows: 0.01–0.20, none to slight; 0.21–0.40, weak; 0.41–0.60, moderate; 0.61–0.80, substantial; and 0.81–1.00, almost perfect. All analyses were performed at a two-sided 5% significance level using SAS software (version 9.4; SAS Institute, Inc., Cary, NC).

Results

Patient population

Of 106 children who underwent chest CT for evaluation of a congenital lung lesion, 23 were excluded: 21 for lack of pathologic confirmation and 2 for subdiaphragmatic sequestration. The final cohort comprised 83 children—52 male (62.7%) and 31 female (37.3%)—with a mean age of 3 months (range, birth to 11 months). Sixty-six children (79.5%) were White, 8 (9.6%) Black or African American, 7 (8.4%) Hispanic or Latino, 1 (1.2%) Asian, and 1 (1.2%) of another race. Ten children (12.0%) were born prematurely, with a mean gestational age of 35 weeks (range, 31 weeks to 36 weeks and 6 days). Thirteen children (15.7%) had one or more associated congenital anomalies: cardiovascular abnormalities (n = 6), renal anomalies (n = 4), chromosomal abnormalities (n = 3), musculoskeletal anomalies (n = 2), congenital diaphragmatic hernia (n = 2), sacral teratoma (n = 1), and myelomeningocele (n = 1).

Pathology of congenital lung lesions

Ten children (10/83, 12.0%) had higher-risk pathology: CPAM type 1 in 8 (8/83, 9.6%) and type I PPB in 2 (2/83, 2.4%). Seventy-three children (73/83, 88.0%) had low-risk pathology. This group included isolated CPAM type 2 in 36 children (36/83, 43.4%), isolated sequestration in 18 children (18/83, 21.7%), and sequestration associated with CPAM type 2 (hybrid lesion) in 15 children (15/83, 18.1%). Four children (4/83, 4.8%) had other pathology: lymphangiectasis (n = 2), bronchogenic cyst (n = 1), and gastric duplication cyst (n = 1).

Imaging characteristics of the lung lesions on chest CT and correlation with pathology

Given the extended study period, CT acquisition techniques varied across vendors and evolving protocols. All examinations were performed with intravenous contrast, except for two. The average size of the lung lesions was 4.4 cm. There was no significant difference in size between the low-risk (average, 4.4 cm; range, 1.5–8.8 cm) and higher-risk categories (average, 5.1 cm; range, 3.2–11.0 cm; p = 0.32). The left lower lobe was the most common location (Table 1), observed in 40 patients (48.2%) by radiologist 1 and 41 patients (49.4%) by

radiologist 2. Lesion location differed significantly between risk categories ($p < 0.001$); left upper lobe involvement was disproportionately represented in the higher-risk group (50.0% for radiologist 1, 44.4% for radiologist 2).

Table 1
Distribution of lesions by pulmonary lobe in 83 patients.

Location, n (%)	Radiologist 1	Radiologist 2
Right upper lobe (RUL)	10 (12.0%)	13 (15.7%)
Right middle lobe (RML)	6 (7.2%)	7 (8.4%)
Right lower lobe (RLL)	26 (31.3%)	26 (31.3%)
Left upper lobe (LUL)	8 (9.6%)	9 (10.8%)
Lingula	0 (0%)	3 (3.6%)
Left lower lobe (LLL)	40 (48.2%)	41 (49.4%)
Multilobar involvement	7 (8.4%)	14 (16.9%)
Because some lesions involved multiple lobes, percentages exceed 100%.		
RUL, right upper lobe; RML, right middle lobe; RLL, right lower lobe; LUL, left upper lobe; LLL, left lower lobe.		

Cystic changes were identified in 40 cases (48.2%) and 42 cases (50.6%) by radiologists 1 and 2, respectively. Among lesions with cystic changes, most demonstrated simple cystic morphology (27 cases [67.5%] and 34 cases [81.0%], respectively) and most contained multiple cysts (33 cases [82.5%] and 34 cases [81.0%], respectively). A systemic feeding artery was identified in 34 studies (41.0%) and 25 studies (30.1%) by radiologists 1 and 2, respectively.

Table 2 summarizes the association between individual imaging characteristics and higher-risk pathology. Features associated with increased risk included mediastinal shift and the presence of cysts, particularly complex and solitary cysts. In contrast, hyperinflation (Fig. 1) and the presence of a systemic feeding artery (Fig. 2) were not associated with higher-risk pathology, and none of the patients without cystic lesions had higher-risk pathology. One patient with CPAM type 1 had associated hyperinflation, likely reflecting bronchial obstruction by a large cyst, as identified by radiologist 1 (Fig. 3).

Table 2
Characterization of lung lesions in 83 patients and relative risk* (95% confidence interval) for higher-risk pathology (CPAM Type 1 or pleuropulmonary blastoma).

Radiologist 1				Radiologist 2		
Characteristic	n (%)	p-value	Relative Risk (95% CI)	n (%)	p-value	Relative Risk (95% CI)
Mediastinal shift	15 (18.1%)	0.005	4.53 (1.50–13.71)	20 (24.1%)	0.041	3.15 (1.01–9.78)
Hyperinflation	36 (43.4%)	0.003	0/36 vs. 10/47	46 (55.4%)	0.002	0.09 (0.01–0.67)
Atretic bronchial stump	8 (9.6%)	0.271	0/8 vs. 10/75	16 (19.3%)	0.099	0/16 vs. 10/67
Feeding systemic artery	34 (41.0%)	0.034	0.16 (0.02–1.21)	25 (30.1%)	0.027	0/25 vs. 10/58
Diaphragmatic hernia	9 (10.8%)	0.240	0/9 vs. 10/74	5 (6.0%)	0.393	0/5 vs. 10/78
Cystic lesion	40 (48.2%)	< 0.001	10/40 vs. 0/43	42 (50.6%)	0.001	10/42 vs. 0/41
Solitary cyst	7 (17.5%)	< 0.001	7.24 (2.66–19.69)	8 (19.0%)	0.236	2.34 (0.60–9.20)
Multiple cysts	33 (82.5%)	0.163	2.27 (0.69–7.44)	34 (81.0%)	0.007	5.76 (1.30–25.49)
Simple cyst(s)	27 (67.5%)	0.367	0.52 (0.12–2.28)	34 (81.0%)	0.452	0.62 (0.17–2.22)
Complex cyst(s)	13 (32.5%)	< 0.001	21.54 (5.14–90.18)	19 (45.2%)	< 0.001	30.32 (4.10–224.35)
Septated	2 (5.0%)	0.095	4.50 (0.99–20.51)	13 (31.0%)	< 0.001	12.56 (3.72–42.39)
Dominant cyst	3 (7.5%)	< 0.001	11.43 (5.63–23.19)	9 (21.4%)	< 0.001	19.19 (6.00–61.33)
Solid components	8 (20.0%)	0.001	6.25 (2.22–17.58)	5 (11.9%)	0.001	6.69 (2.45–18.28)
Largest cyst size†	≥ 2.4 cm	< 0.001	44.4 (6.10–323)	≥ 2.2 cm	< 0.001	10/15 vs. 0/68
* Relative risk is not shown when the probability of higher-risk pathology is 0% for patients with or without the characteristic; frequencies (patients with vs. without the characteristic) are shown instead.						
† Largest cyst size most strongly associated with higher-risk pathology, derived using classification tree analysis.						

The size of the largest cyst was also associated with higher-risk pathology (mean, 3.7 cm for higher-risk and 1.5 cm for lower-risk lesions per radiologist 1; 3.6 cm and 1.4 cm, respectively, per radiologist 2). However, cyst size alone could not reliably distinguish lower-risk from higher-risk lesions. Radiologist 1 identified cysts > 2 cm in 6

of 73 patients (8.2%) with lower-risk lesions (CPAM type 2, n = 4; hybrid lesion, n = 1; bronchogenic cyst, n = 1) and cysts ≤ 2 cm in 1 of 10 patients (10%) with CPAM type 1. Radiologist 2 identified cysts > 2 cm in 5 of 73 patients (6.8%) with lower-risk lesions (CPAM type 2, n = 4; bronchogenic cyst, n = 1) and no higher-risk lesions with cysts ≤ 2 cm (Figs. 4 and 5). Interobserver agreement for cyst-size measurement was high (intraclass correlation coefficient, 0.99; within-patient standard deviation, 0.21 cm).

Using CT criteria that defined low-risk cystic lesions as those with no cysts, a simple cyst ≤ 2 cm, or any cyst with an identified systemic feeding artery, 65 patients (78.3%, 65/83) were classified as low risk by radiologist 1 and 68 patients (81.9%, 68/83) by radiologist 2; all had low-risk pathology. With these criteria, sensitivity was 100% for both radiologists, specificity was 89.2% (radiologist 1) and 93.1% (radiologist 2), PPV was 55.6% and 66.7%, NPV was 100% for both, and overall accuracy was 90.5% and 94.0% for radiologists 1 and 2, respectively.

Risk categories of congenital lung lesions assigned by radiologists compared with pathology

Table 3 compares the low-risk and higher-risk diagnostic categories assigned by radiologists 1 and 2 with the corresponding pathologic diagnoses and risk categories. Sensitivity was 100% for both radiologists; specificity was 89.2% and 93.1%; PPV was 56.3% and 69.2%; NPV was 100% for both; and overall accuracy was 90.5% and 94.0% for radiologists 1 and 2, respectively. Interobserver agreement on risk level was almost perfect (98.4% agreement; $\kappa = 0.95$; 95% CI, 0.85–1.00) for the 63 lesions rated with moderate or high confidence.

Table 3

Diagnosis with moderate or high confidence of congenital lung lesions by radiologists compared with pathology

	Pathology diagnosis							
	Low risk					Higher risk		
Radiologists' diagnosis	Sum	Sequestration	CPAM type 2	Lymphangiectasis	DC	Sum	CPAM type 1	PPB
Radiologist 1								
Low risk	58 (100%)	29	26	2	1	0	0	0
Higher risk	7 (43.7%)	0	6	0	1	9 (56.3%)	7	2
Indeterminate	8 (88.9%)	4	4	0	0	1 (11.1%)	1	0
Radiologist 2								
Low risk	54 (100%)	25	26	2	1	0	0	0
Higher risk	4 (30.8%)	0	3	0	1	9 (69.2%)	7	2
Indeterminate	15 (93.8%)	8	7	0	0	1 (6.3%)	1	0
CPAM congenital pulmonary airway malformation, DC duplication cyst, PPB pleuropulmonary blastoma type 1								
Values in the Sum columns are the number (row percentage) of lesions in each pathology risk group; remaining columns give the count of each specific diagnosis								

When low-confidence classifications were included, the two radiologists agreed on risk classification in 67 of 83 patients (80.7%); consensus review resolved the discrepancies in the remaining 16 patients. After consensus, higher-risk lesions were diagnosed in 18 patients, which included all 10 higher-risk pathologies. Low-risk lesions were diagnosed in 60 patients, all of whom had low-risk pathology. An indeterminate-risk classification was assigned in 5 patients, all of whom had low-risk pathology. For the radiologists' consensus risk-category assignment, sensitivity was 100%, specificity was 88.2%, PPV was 59.2%, NPV was 100%, and overall accuracy was 89.6%.

Discussion

This study demonstrates that radiologists can reliably distinguish low-risk from higher-risk (CPAM type 1 and type I PPB) congenital lung lesions on chest CT, with high sensitivity (100%), high NPV (100%), and almost-perfect interobserver agreement (98.4%; $\kappa = 0.95$). These findings support CT-based risk stratification to guide management in selected asymptomatic children, allowing imaging surveillance as an alternative to routine surgical resection.

There are several reasons to classify CPAM type 1 within the higher-risk category. First, chest CT cannot reliably differentiate CPAM type 1 from type I PPB, as both most often present as macrocysts^{21,23,24}. Second, compared with other congenital lung lesions, CPAM type 1 contains mucinous cell clusters in up to 80% of cases, frequently harbors KRAS mutations, and may transform into mucinous adenocarcinoma in up to 1% of cases^{7,9,25}. Although this risk is substantially lower than that of type I PPB which progresses to the aggressive type II or III in approximately 10% of cases¹⁹ and represents the predominant form detected on prenatal US and in infancy^{19,26}, it is not negligible. Third, CPAM type 1 carries a higher risk of complicating infection^{11,12,27,28}. This last point is one of the main arguments for early resection of congenital lung lesions, as later surgery for symptomatic lesions, including infected lesions, is associated with increased complications and longer hospital stay^{19,29–31}.

Although cyst size is a key morphologic feature for distinguishing higher-risk lesions from CPAM type 2, it cannot be used in isolation, given the overlap. Prior studies have shown that up to 20% of CPAM type 1 lesions have a maximal cyst diameter < 2 cm³². In our cohort, one CPAM type 1 lesion (10%) had a maximal cyst diameter < 2 cm per radiologist 1 and a borderline measurement of 2.2 cm per radiologist 2. Data from the PPB registry indicate that although most PPBs exceed 5 cm²⁶, the lower size range for type I and type Ir is 2 cm and 0.4 cm, respectively¹⁹. Evaluation of complex cyst features is therefore important for differentiating among the types of congenital lung lesion. Registry data indicate that 96% of type I PPBs are multiseptated¹⁹. In our study, complex cysts had relative risks of 21.5 and 30.3 (radiologists 1 and 2, respectively) for CPAM type 1 or type I PPB.

In our cohort, lesions were categorized as higher risk if they demonstrated a solitary cyst, a complex cyst, or a cyst with a maximal diameter > 2 cm, corresponding to CPAM type 1 or cystic PPB. Low-risk lesions included those with predominant hyperinflation, with or without cysts < 2 cm (CPAM type 2), and lesions with an identified systemic feeding artery (pulmonary sequestration).

On analysis of the imaging features, we found that hyperinflation and a systemic feeding artery were associated with low-risk lesions. In addition, all cases with an atretic bronchial stump or diaphragmatic hernia were low risk, although the numbers were too small to reach statistical significance. Mediastinal shift, complex cysts, and larger cyst size were all associated with higher-risk lesions. Applying the criteria of absent cysts, simple cysts ≤ 2 cm, or cystic lesions with a systemic feeding artery correctly classified all low-risk lesions and encompassed most patients (78.3% and 81.9% for radiologists 1 and 2, respectively), with 100% sensitivity and NPV.

Other authors have proposed CT-based criteria including hyperinflation, systemic feeding vessels, multifocal or bilateral disease, mediastinal shift, and complex cysts—combined with DICER1 mutation screening to improve risk stratification². Notably, one of our two PPB cases lacked any of these higher-risk imaging features. In addition, because localized hyperinflation can result from bronchial obstruction by a cystic lesion, as seen in one of our CPAM type 1 lesions, it cannot be used as an absolute indicator of a low-risk lesion. We therefore suggest using the criteria applied in our study, a lesion with predominant hyperinflation associated with cysts < 2 cm.

No uniform approach exists for the management of asymptomatic pulmonary sequestration and congenital cystic lung lesions; superimposed infection, which increases surgical complications, and, to a lesser degree, malignancy risk are the primary indications for surgery^{1,16,29,30}. The recurring argument for elective resection is that operating while the patient is asymptomatic carries a lower surgical risk than waiting for symptoms to develop and then operating^{29,30}. The counterargument for conservative management is that the absolute

complication rate of elective surgery for asymptomatic lung lesions was found in a meta-analysis to be 16% (63/404) for thoracoscopic operations and 18% (87/483) for open thoracotomy³³ may outweigh the risk posed by infected cysts, as most lesions never become symptomatic⁴.

A longitudinal study following 68 children with congenital cystic lung lesions over a median of 9.9 years found that the risk of recurrent infection was less than 10%, most common CPAM type 1, and decreased after the second year of life, with no cases of malignancy identified³⁴. Using a decision-analytic model, Yang et al. analyzed quality-adjusted life-years for two hypothetical asymptomatic 6-month-old children with congenital lung lesions—one with a large lesion occupying more than 50% of the involved lobe and one with a small lesion occupying less than 50%, and found that expectant management yielded a small gain (0.09 and 0.15 quality-adjusted life-years, respectively) relative to surgery⁴. In addition, some pulmonary sequestrations and cystic lung lesions decrease in size or involute^{34–38}, with a low risk of subsequent enlargement or complication³⁷.

The optimal follow-up duration and imaging strategy for conservatively managed patients remain undefined. Periodic chest radiographs at 6- to 12-month intervals, while less sensitive than CT, may be sufficient to exclude significant progression, with the advantages of lower radiation exposure and reduced cost. Because complications requiring surgical intervention beyond 5 years of age are uncommon³⁴, with only rare cases reported in older children^{12,13}, imaging follow-up beyond 5 years may not be necessary. Most late-presenting CPAM cases in adults are CPAM type 1¹⁰. Our approach of advocating resection of type 1 lesions regardless of size is therefore likely to reduce the long-term risk of infection and malignant transformation.

This study has several limitations. Its retrospective, single-institution design and the small number of higher-risk lesions (8 CPAM type 1 and 2 PPB) limit generalizability. Multicenter studies with larger cohorts are needed to validate CT-based risk stratification and to assess diagnostic performance using receiver operating characteristic analysis. Finally, long-term outcomes, including the risk of recurrent infection or malignant transformation in conservatively managed lesions, were not evaluated.

In conclusion, radiologists can reliably classify congenital lung lesions as higher risk (CPAM type 1 and PPB) versus low risk (CPAM type 2 and pulmonary sequestration) using chest CT, with high sensitivity, high NPV, and almost perfect interobserver agreement. Higher-risk lesions are unlikely when a lung lesion demonstrates no cysts, simple cysts ≤ 2 cm, or cysts with a systemic feeding artery. In asymptomatic children who meet these low-risk criteria, conservative imaging surveillance may be considered as an alternative to surgical resection. Future multicenter studies with larger cohorts are needed to validate these findings.

Declarations

Conflicts of Interest:

None

Competing interests:

The authors have no competing interests to declare that are relevant to the content of this article.

Ethics approval:

This retrospective study was approved by the Indiana University Institutional Review Board, Indianapolis, Indiana (IRB #2024 – 00170), in accordance with the ethical standards of the 1964 Declaration of Helsinki and its later amendments.

Informed consent:

Informed consent was waived by the Indiana University Institutional Review Board in view of the retrospective nature of the study.

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Author Contribution

B.K. conceptualized and designed the study and drafted the initial manuscript. S.C. performed image review and revised the study. M.C. performed image review and revised the study. I.S. assisted with clinical data collection and revised the study. E.A. revised pathology slides and revised the study. M.M. conceptualized the study and revised the study. B.B. conceptualized the study and revised the study. J.C. assisted with clinical data collection and revised the study. G.E. assisted in the statistical analysis and revised the study. B.G. assisted in conceptualizing and designing the study and revised the study. All authors reviewed and approved the final manuscript.

Data availability:

The data are not publicly available in order to preserve individuals' privacy.

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Figures

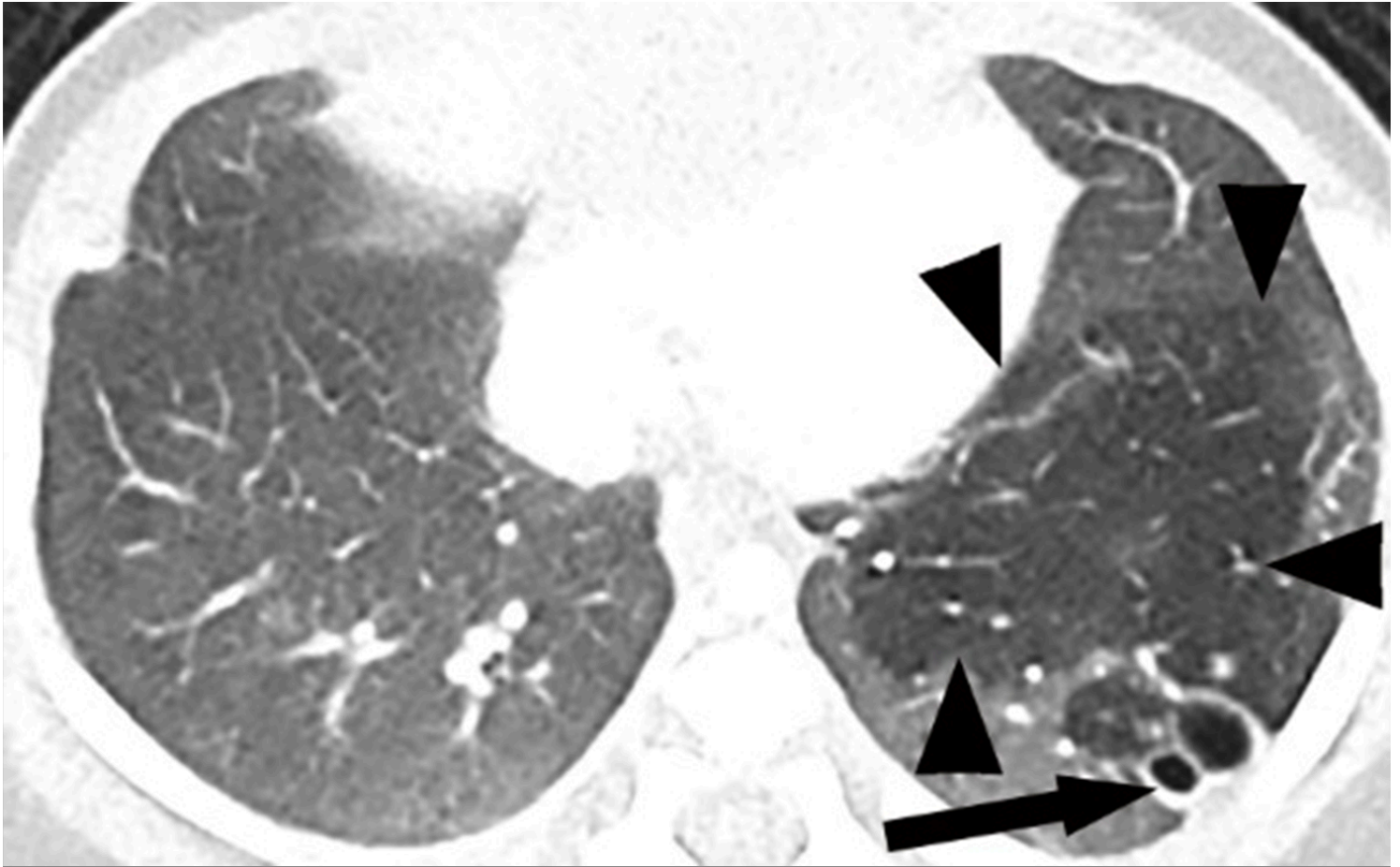


Figure 1

CPAM type 2 with predominantly emphysematous changes and small cysts in a 3-month-old male. Axial chest CT demonstrates focal emphysematous changes (arrowheads) and small cysts (arrow). This was classified as low-risk lesion by both radiologists.

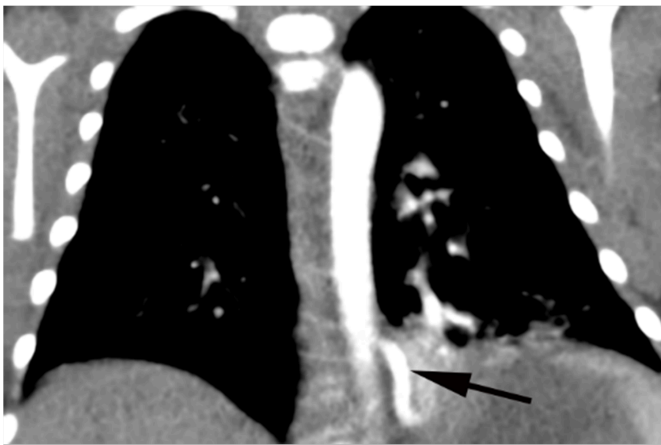


Figure 2

CPAM type 1 in a 3-month-old female with large cysts and adjacent emphysematous change. A. Sagittal chest CT shows multiple large cysts with adjacent emphysematous changes (arrows), likely related to bronchial obstruction by a large cyst. B. Axial chest CT shows focal emphysematous change in the lingula (arrow). This lesion was classified as higher risk by both radiologists.

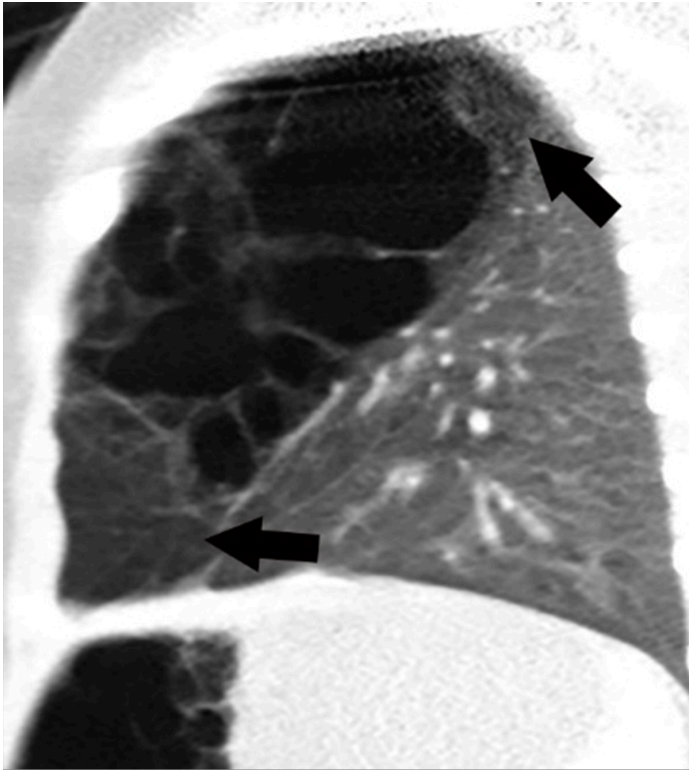


Figure 3

Hybrid congenital lung lesion with small cysts, solid component and systemic feeding vessel in a 5-month-old female. A. Axial chest CT image demonstrates multiple small cysts associated with a solid component (arrow). B. Coronal CT image shows a systemic feeding artery (arrow). C. Three-dimensional CT reconstruction demonstrates the systemic feeding artery (arrow) and draining pulmonary veins (arrowheads). This was classified as low-risk lesion by both radiologists.

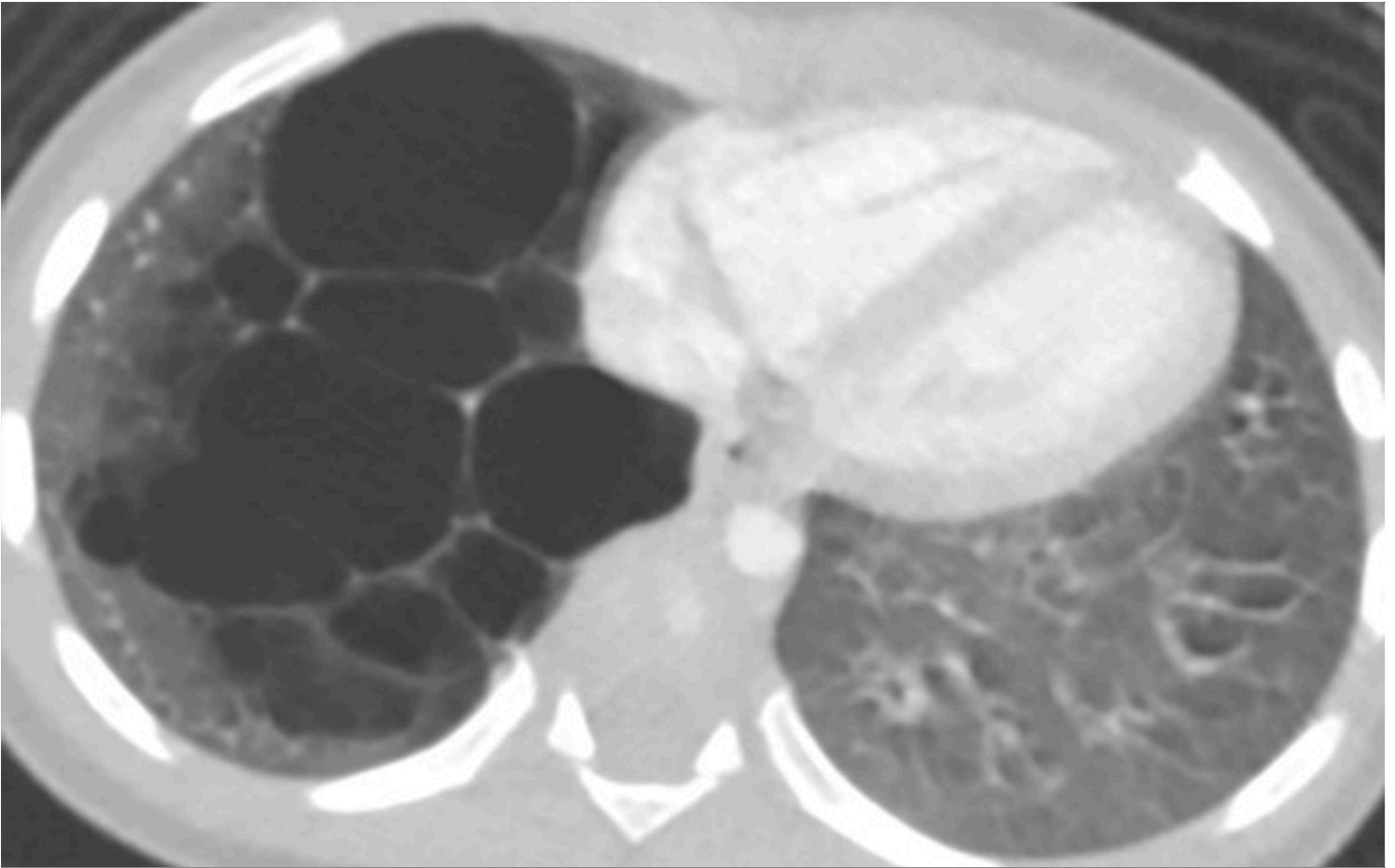


Figure 4

CPAM Type 2 with large cysts in 11-month-old girl. Axial chest CT image demonstrates multiple large cysts in the right lower lobe. This was falsely classified as higher risk lesion by both radiologists.

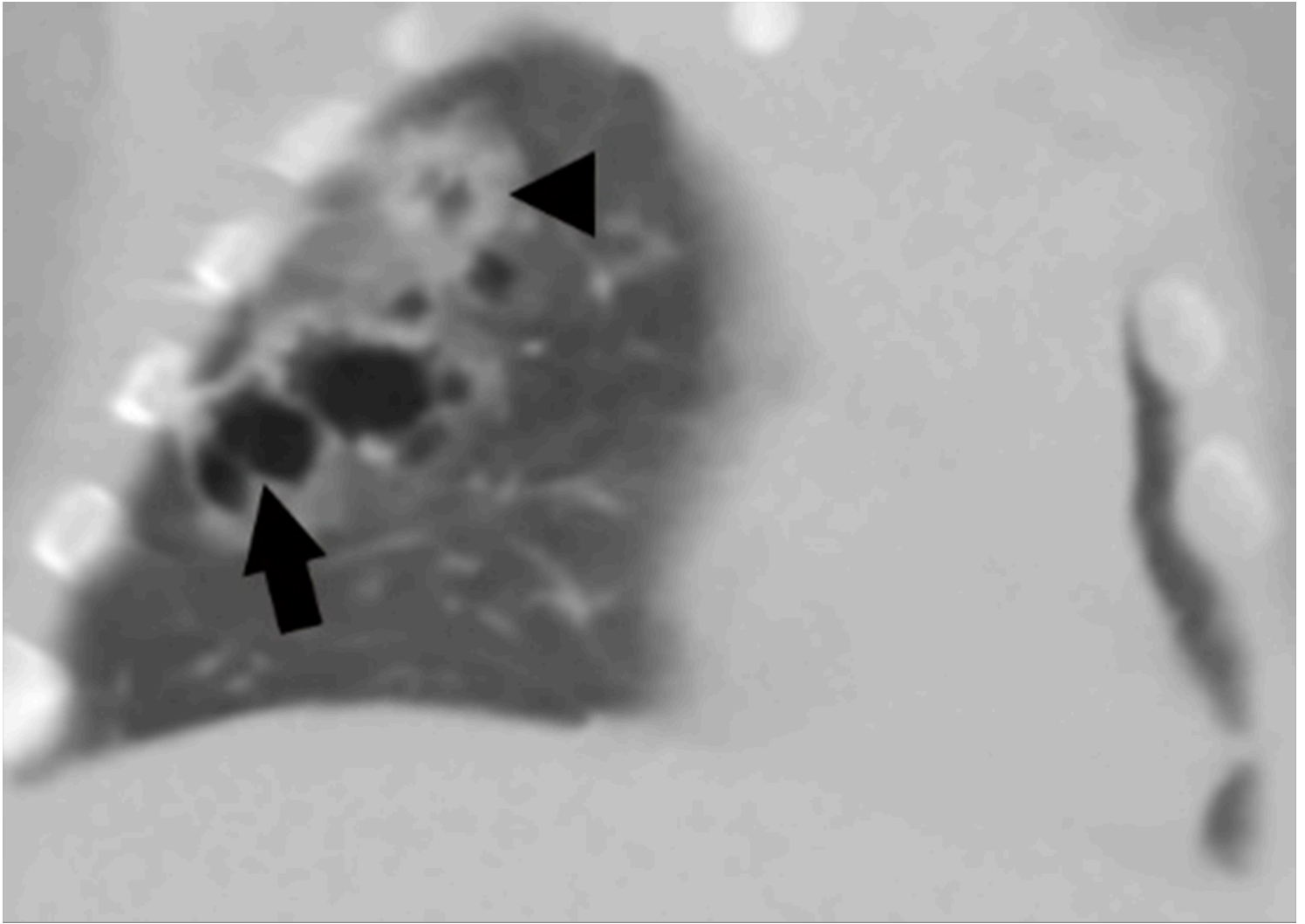


Figure 5

CPAM type 1 in a 3-month-old male with multiple small complex cysts; the largest cyst measured 1.7 cm by radiologist 1 and 2.2 cm by radiologist 2. Coronal chest CT demonstrates a conglomerate of small septated cysts (arrow) with a thick wall (arrowhead) and marked leftward mediastinal shift. Both complex cysts and mediastinal shift were associated with higher risk lesions. One radiologist classified the lesion as higher risk and the other as indeterminate risk.

Supplementary Files

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