REVIEW ARTICLE



Role of Computed Tomography in Pediatric Chest Conditions

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Abstract CT is the preferred cross-sectional imaging modality for detailed evaluation of anatomy and pathology of the lung and tracheobronchial tree, and plays a complimentary role in the evaluation of certain chest wall, mediastinal, and cardiac abnormalities. The article provides an overview of indications and different types of CT chest, findings in common clinical conditions, and briefly touches upon the role of each team member in optimizing and thus reducing radiation dose.

Keywords Computed tomography · CT · Pediatric · Chest

Introduction

Although the plain radiograph is the first imaging modality of choice to evaluate suspected chest conditions, computed tomography (CT) is considered the most valuable modality to accurately evaluate airway, cardiovascular and mediastinal abnormalities [1–5], as well as the lung parenchyma. Sometimes, it may be necessary to use other imaging modalities like ultrasound and MRI for further characterization of the nature and extent of the disease. Table 1 summarizes the indications and role of these modalities.

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Multidetector CT

Currently, low radiation protocol multidetector CT (MDCT) is considered to be the ideal way to assess the lung parenchyma, central airway, mediastinal and cardiovascular abnormalities in children [3]. With MDCT, thin sub millimeter CT sections are acquired to obtain isotropic volumetric data, which can be subjected to various reformations like multiplanar reformations (in axial, coronal, sagittal or any oblique plane), volume rendered 3D reconstructions, maximum and minimum intensity projections, to detect, characterize and display the abnormalities, aiding in pre-surgical planning and patient management. Virtual bronchoscopy can also be performed.

Faster exam techniques result in improved image quality by reducing motion and respiratory artefacts and enable depiction of small structures and vessels [6] and also reduced requirement of sedation and anesthesia.

A CT exam results in increased radiation exposure which has associated potential risks in the pediatric population that cannot be ignored [7–9]. Table 2 highlights the radiation doses of chest radiographs and MDCT [10, 11]. Radiation protection has three identifiable tenets *viz.* justification, optimization and dose limitation [12].

Justification

Eliminating unnecessary and inappropriate referrals is the most effective method of reducing radiation [13, 14]. Strict referral criteria can be developed locally by consensus, or existing published guidelines can be followed [15–17]. All requests should be justified by the referring clinician as well as the radiologist. Available imaging should be reviewed and non-ionizing radiation modalities should be considered as an alternative, wherever appropriate.

Table 1	Role of different in	naging modalities	in evaluating pediatric	chest conditions
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Modality	Uses	Advantages	Disadvantages
Ultrasound	 Assessing synpneumonic and parapneumonic pleural effusions (quantifying, assessing extent and nature of fluid and loculations) Small chest wall masses (characterizing) 	No radiationEasily available	 Operator dependent Cannot visualize deep seated lesions, lung parenchyma, bone abnormalities
CT scan	 Chest wall Chest wall deformities Chest wall masses (to characterize, assess extent and also for bone involvement) Chest wall injury Lung Persistent or recurrent infections (to look for complications, underlying anomalies as causes) Neoplasms (primary lung masses and also to look for metastases in extrapulmonary malignancy) Diffuse or interstitial lung disease [<i>e.g.</i>, Cystic fibrosis, interstitial lung disease [<i>ILD</i>) associated with connective tissue disease] Congenital anomalies [like bronchogenic cyst, congenital pulmonary airway malformation (CPAM), congenital lobar emphysema] Trauma (lung contusion or lacertaion) Mediastinum Congenital anomalies (like bronchogenic cyst) Neoplasms (mainly for assessing anterior mediastinal masses like thymic lesions, lymphoma and germ cell tumors) Congenital and acquired exrtacardiac vascular abnerosition 	 Easily available Short examination times (less need for anesthesia or sedation) Multiplanar reconstructions possible Modality of choice for assessing lung parenchymal changes Modality of choice for assessing bone abnormality 	 Exposure to ionizing radiation May require exposure to iodinated contrast which can be potentially nephrotoxic
MRI	 Chest wall Spinal deformities (to look for associated cord abnormalities) Chest wall masses (to characterize, assess extent. CT scan may be better to assess the bone involvement) Lung Currently, CT is better in assessing the lung parenchymal abnormalities. Studies show role of MRI in chronic lung parenchymal abnormalities like cystic fibrosis, to avoid radiation from multiple follow up CT scans. Mediastinum Neoplasms (mainly for assessing posterior mediastinal masses like neurogenic tumors and neuroenteric cysts. Spinal canal extension and changes in the spinal cord and nerve roots are best assessed by MRI) MRA can be tried in exrtacardiac vascular abnormalities, again, when multiple follow up examinations may be needed 	 Safe examination as there is no exposure to ionising radiation and so, is better modality when repeated follow ups are required. Modality of choice for assessing neural foraminae, spinal canal, spinal cord. 	 Longer examination time (more requirement of anesthesia / sedation). Other than when 3D sequences are obtained, multiplanar reconstructions are not optimal. Not ideal for assessing lung parenchymal and bone abnormality.

Optimization and Dose Limitation

It is important to tailor the protocol such that only the part of the body that is required to answer the clinical question be covered in the examination. The weakness or tendency to image adjacent structures 'while the child is in the scanner' should be avoided. Children-specific parameters based on either age, weight, or body size are chosen either inherent within the scanners, or by following existing guidelines [18–22].

 Table 2
 Comparison of radiation dose from chest radiograph and CT chest and abdomen

Examination	Typical effective dose (mSv)	Equivalent number of chest radiographs	Equivalent length of background exposure
Chest radiograph	0.02	1	2.4 d
CT chest	8	400	3.6 у
CT abdomen or pelvis	10	500	4.5 y

Table 4Endobronchiallesions in children [26]

ial 5]	Commoner lesions	Foreign bodies Carcinoid
		Mucoepidermoid carcinoma
		Adenoid cystic carcinoma
	Rare lesions	Myofibroblastic tumor
		Hamartomas
		Pulmonary chondroma
		Inflammatory polyps

Technique

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CT of the chest can be done technically in different ways depending on the clinical indication. Table 3 shows the two main types of CT chest - with (contrast enhanced CT or CECT) or without (plain / non-contrast CT or NCCT) administering intravenous iodinated contrast agents. Other variations like CT pulmonary angiogram where the contrast injection is timed to get maximum enhancement of the pulmonary arteries, cardiac CT where the contrast is in the cardiac chambers, CT angiogram of the aorta can also be done according to the clinical indication.

COT C 1

High Resolution CT Thorax (HRCT Thorax)

For HRCT, classically, sequential thin section (1-1.5 mm) images at 10 or 20 mm intervals are obtained with excellent spatial resolution of the lung parenchyma achieved by using high spatial frequency reconstruction algorithm.

In the era of MDCT, HRCT images can be reconstructed in all patients from the volumetric helical data acquired during the plain or contrast enhanced MDCT and this technique is followed in most centres now.

lable 3	Main	types	ofCl	ls of	chest	

Type of CT	Indications	Advantages	Disadvantages	
Non-contrast CT or NCCT 1. When predominantly lung parenchymal abnormality is suspected [24], like: • Interstitial lung disease • Bronchiectasis • Cystic fibrosis • Bronchopulmonary dysplasia • Severe asthma • Bronchiolitis obliterans 2. To assess bony chest wall abnormalities 3. When contrast is contraindicated (allergy, renal impairment etc.)		No use of iodinated contrast material.	Cannot optimally assess mediastinal lesions and lymphadenopathy.	
Contrast enhanced CT or CECT	 (anteg), this explanation of the mediastinal abnormalities need to be assessed, like: Known or suspected infection <i>e.g.</i>, complicated pneumonia, abscess, tuberculosis, empyema Lung, anterior or middle mediastinal, chest wall masses Suspected congenital abnormalities like sequestration Vascular disorders Trauma which is inadequately assessed by CXR. 	Mediastinal and vascular structures are well assessed in addition to the lung parenchyma.	Use of iodinated contrast material with its potential risk of nephrotoxicity and requirement of more sedation.	

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	Role	Limitations
MDCT with virtual bronchoscopy	 Accurate noninvasive localization of bronchial obstruction, thus aiding fibreoptic bronchoscopy Can see 'beyond' the level of obstruction In addition to the endoluminal component, can see lesions in bronchial wall, extrinsic lesions compressing bronchus. 	May not be able to diagnose accurate nature of the pathology, thus, biopsy and histopathology may be needed.
Rigid or flexible bronschoscopy	Can do a diagnostic biopsy of the lesion or therapeutical removal of foreign body or debulking of tumor.	 Invasive Can mainly see the endobronchial component and cannot see 'beyond' the narrowing

Table 5 Role of CT and fibreoptic bronschoscopy in suspected endobronchial lesion

However, acquiring contiguous helical volumetric data would result in 5–10 times more radiation than the sequential HRCT with inter slice gap [23].

Routine acquisition of pre-contrast CT prior to the contrast enhanced images is not recommended as it does not add any extra value [24].

Commonly encountered clinical conditions where CT chest would be done, common causes for each clinical condition and their imaging are briefly addressed. For a more vast coverage and further information, the reader is encouraged to refer to recent reference text books.

Common Clinical Conditions

- 1. Recurrent or chronic infection (including specific infections).
- 2. Unilateral hyperlucent hemithorax.
- 3. Lung mass.

Fig. 1 Endobronchial mucoepidermoid tumor. CXR (a), contrast enhanced axial CT section in mediastinal window (b) and coronal CT section in lung window at the level of the carina (c) in a 14-y-old boy with recurrent lower respiratory infection (LRI) for 1 y, show, an obstructing endobronchial mass in the left main bronchus (*arrows*) with collapse of the left lung (*multiple thin arrows* in b)

- 4. Mediastinal mass.
- 5. Chest wall mass.
- Cough and breathlessness or suspected diffuse parenchymal or interstitial lung disease.
- 7. Miscellaneous

Recurrent or Chronic Infection

Radiographs remain the initial modality for diagnosis. CT is used in cases of non responsiveness to treatment, to evaluate complications, and underlying congenital abnormalities.

Endobronchial Lesions (Table 4) [25]

They can present with recurrent infections or persistent wheeze. When recurrent infections occur in the same



Fig. 2 Endobronchial radiolucent foreign body. A 14-yold with recurrent on and off fever and respiratory symptoms for 4 y. CXR (a) shows collapse of right middle and lower lobe with bronchiectatic changes (*arrows*). Axial CT chest in lung window (b) shows an ovoid foreign body, a peanut (*curved black arrow*) occluding the bronchus intermedius and CT sections at a lower level (c) shows the collapsed right middle and lower lobe with bronchiectatic changes



location or when there is persistent opacity on CXR in between the episodes of infection, either a congenital abnormality in that location or an obstruction to the supplying bronchus should be suspected [26]. CECT would be the best imaging modality to further evaluate such patients. Virtual bronchoscopy images can also be reformatted with the MDCT volumetric data. Role of CT and fibreoptic bronschoscopy in such situations is discussed in Table 5 [27] (Fig. 1). Brightly enhancing lesions should suggest carcinoid tumors.

Endobronchial Foreign Body

Depending on the degree of bronchial obstruction, air trapping or atelectasis can be seen distal to the foreign body and recurrent infections can develop. CT can help in localizing the foreign body (can detect even radiolucent foreign bodies which will not be visible on CXR (Fig. 2) and also in assessing any complications in the distal lung (Fig. S1) [28].

Sequestration (Table 6)

It consists of non-functioning lung tissue which does not communicate with the bronchial tree and receives systemic arterial supply (Fig. S2).

Immunocompromised Children

CT is useful in looking for complications in immunocompromised patients with lung infections (Fig. S3). CT may be indicated in immunocompromised patients, even in the absence of chest radiographic abnormalities, for early detection of suspected opportunistic infections [29].

Table 6 Types of sequestration

	Intralobar	Extralobar
Pleura	No separate pleural covering	Has separate pleural covering
Tracheo-bronchial tree	No communication	No communication
Arterial supply	Systemic	Systemic
Venous drainage	Pulmonary veins	Systemic veins
Associated anomalies	Less common	More common; include congenital diaphragmatic hernia, vertebral anomalies, congenital heart disease, pulmonary hypoplasia, and colonic duplication



Fig. 3 Staphylococcal pneumonia. CT thorax in lung (a) and mediastinal window (b) show a large area of consolidation in the right lower lobe (*straight longer arrows*), pleural effusion (*triple arrows*) and a cystic airspace medially indicating development of a pneumatocele

Immunodeficiency should be suspected in patients with recurrent infections with unusual organisms or atypically severe, complicated, persistent or recurrent infections with usual organisms.

Chest findings are present in upto 60 % of patients who have primary immunodeficiency [30]. Imaging, including CT chest plays an important role in identifying the lung findings, may aid in diagnosis, helps in risk stratification, prognostication and monitoring response to therapy [31].

Tracheo-esophageal Fistula

Although CT is not the modality of choice to diagnose tracheo-esophageal fistula (Esophagogram is the modality of choice), this finding may sometimes be unexpectedly detected in children with recurrent infection (Fig. S4).

Bacterial Infections (Fig. 3)

Lobar or patchy areas of consolidations are the common findings. Thin walled rounded air filled spaces called pneumatoceles [32] may develop in some pneumonias, commonly in Staphylococcal pneumonia. They can appear during the 1stwk of infection and usually spontaneously resolve by 4–6 wk.

Tuberculosis

a) Primary tuberculosis (Figs. 4 and S5): Common findings include parenchymal consolidation (common in middle

Fig. 4 Primary tuberculosis. Chest radiograph (a) shows parenchymal consolidation in right upper and mid zones with hilar lymphadenopathy (*black arrows*). Contrast CT thorax of different patients (**b**, **c** and **d**) show pleural effusion and thickening indicating empyema (*curved white arrows*), parenchymal consolidation (*black arrow*) and enlarged lymph nodes with central hypodense attenuation suggestive of necrosis (*white arrows*)



Fig. 5 Ruptured hydatid cyst. CT thorax shows collapsed membranes (water lily sign) within an air filled cavity (*black arrow*)

and lower lobe), lymphadenopathy, miliary nodules and pleural effusion.

b) Post primary tuberculosis (Figs. S6 and S7): Results from re-infection or reactivation of primary tuberculosis. Imaging findings include parenchymal opacities with cavitation (upper lobe predilection), airway involvement and pleural effusions.

Invasive Fungal Infection

Invasive fungal infections are seen in immunocompromised children. Nodules with surrounding ground glass opacification due to perilesional hemorrhage ('halo sign') can be seen (Fig. S8). Air crescent sign is seen in the recovery phase when neutropenia settles and the infracted tissue retracts (tissue infarction is due to angio-invasion). The imaging features of pulmonary hydatid cyst (Fig. 5) depend on the stage of the hydatid cyst. Cystic lesion (density of -20HU to 10HU) with a well defined wall can be seen in an unruptured cyst. Once the cyst ruptures, crescent of air can be seen between the layers of the wall, floating folded membranes, or small trapped foci of air could be seen.

Unilateral Hyperlucent Hemithorax (Table 7)

Congenital Lobar Emphysema

Here, there is hyperinflation of one or more lobes due to intrinsic or extrinsic bronchial obstruction. Contrast CT is preferred to look for causes of extrinsic obstruction which include foregut cysts (Fig. 6) and vascular anomalies like pulmonary artery sling.

Congenital Pulmonary Airway Malformation or Cystic Adenomatoid Malformation

It occurs due to abnormal bronchoalveolar development and can appear as multiple air filled cysts (Fig. 7) or solid areas depending on the type.

Bronchogenic Cyst

It occurs due to malformation of bronchial tree and can occur in the mediastinum or be intrapulmonary. They can have low or high density fluid as contents or be air filled, if communicating with bronchial tree (Fig. S9).

 Table 7
 Common causes for unilateral hyperlucent hemithorax on chest radiograph

Technical factors	Rotation
Contralateral hemithorax more opaque	Agenesis, hypoplasia, collapse, fibrosis
Abnormality on side of increased lucency	Lung parenchyma Congenital lobar emphysema, Swyer James syndrome, ruptured bronchogenic cyst, Congenital pulmonary airway malformation
	Airways Intrinsic obstruction – foreign body, mucus plugs, mucosal folds In the bronchial wall – maldevelopment of the cartilages, malacia Extrinsic obstruction – vascular rings or mediastinal lesions like foregut cysts
	Vascular Pulmonary artery occlusion like embolism or mass compressing it
	Chest wall Polands syndrome (absent pectoralis major muscle)

Fig. 6 Congenital lobar emphysema. A 4-wk-old infant with breathing difficulty. Chest radiograph (a) shows hyperinflated left hemithorax (arrow) with hyperlucency and mediastinal shift to the right. Plain chest CT in mediastinal (b) and lung window (c) show narrowing of the left main bronchus due to a hypodense lesion (curved arrow) and hyperinflated left lung with increased lucency (arrow in c). There is also a pneumothorax (triple arrows). T2W axial MRI (d) confirms the hyperintense cystic lesion (proven to be bronchogenic cyst on surgery)



Obliterative Bronchiolitis

Swyer James syndrome or Mcleods syndrome is a form of post infectious obliterative bronchiolitis characterised by reduced or normal volume in affected lung or segments which are hyperlucent, with reduced caliber and number of pulmonary vessels (Fig. S10), associated bronchiectasis and bronchial wall thickening. Ipsilateral hilum also appears smaller [33].

Bronchial Atresia

It results from focal stenosis of a bronchus with mucoid impaction distally (forming a branching tubular bronchocele) and hyperinflation of surrounding lung parenchyma (Fig. S11).

Pulmonary Agenesis, Aplasia or Hypoplasia

Patients with lung agenesis (absence of the lung, bronchus, and pulmonary artery) (Fig. S12), aplasia (rudimentary main bronchus) and hypoplasia (hypoplastic bronchus and pulmonary artery with variable amount of lung tissue) will show hyperinflation of the contralateral lung.

They can be associated with other congenital anomalies including vertebral anomalies, cardiovascular defects, anorectal malformations, esophageal atresia, tracheoesophageal fistula, and genitourinary anomalies.

Fig. 7 Congenital pulmonary airway malformation. Chest radiograph (a) shows increased lucency in mid and lower right hemithorax (*short arrows*) with compression and crowding of vessels inferiorly (*long arrow*). Axial CT thorax in lung window (b) shows a large cyst (*) surrounded by multiple smaller cysts (*arrows*)



Fig. 8 Pleuropulmonary blastoma. CXR (a) shows opaque right hemithorax with contralateral mediastinal shift. Axial contrast enhanced CT Chest image demonstrates a large irregular heterogeneously enhancing mass lesion (*arrow*) in the right hemithorax with hyperdense enhancing soft tissue components, infiltrating the mediastinal structures



Lung Mass

Inflammatory Myofibroblastic Tumor (Fig. S13)

Although rare, it is the commonest benign lung tumor in childhood [34]. On CT, it can be a large nodular moderately enhancing peripheral lesion with or without calcification (most common appearance) or an endobronchial mass (2ndmost common appearance).

Metastases (Fig. S14)

They form the most common malignant lung mass in children. Wilms tumor, osteogenic sarcoma, rhabdomyosarcoma, lymphoma and testicular tumours have high propensity for lung metastases. On CT, metastasis are seen as multiple rounded nodules with basal and subpleural predominance. It is a rare pulmonary malignancy. On CT, large heterogenous pleural based hypodense mass with whorls of solid tissue can be seen filling up the hemithorax which may invade the mediastinum and chest wall [35].

Rhabdomyosarcoma (Fig. S15)

Pleuropulmonary Blastoma (Fig. 8)

Rhabdomyosarcoma of the chest wall is more common than primary from pleura or lung. CT findings are nonspecific, with large heterogeneous mass filling up the hemithorax. CT helps in assessing the extent of the lesion and to look for metastases.

Table 8 Common mediastinal masses in children

Anterior mediastinal masses	Middle mediastinal masses	Posterior mediastinal masses
• Lymphoma	• Lymphoma	Ganglion cell tumors:
Germ cell tumors	Bronchogenic cysts	- Neuroblastoma
Thymic masses		- Ganglioneuroblastoma
 Lymphangioma 		- Ganglioneuroma
		Nerve sheath tumors
		• Lymphoma
		Neuroenteric cyst



Fig. 9 Teratoma. Axial contrast enhanced CT Chest image showing a large left anterior mediastinal mass with areas of fat density (*arrows*), calcific foci (*long thin arrow*) and cystic areas (*curved arrows*)

Fig. 10 Bronchogenic cyst. Chest radiograph (a) and contrast enhanced axial CT image (b) showing a well defined cystic mediastinal mass. Non-contrast axial CT in another patient (c) showing a well defined ovoid low density anterior mediastinal bronchogenic cyst (*curved arrow*)



Mediastinal Masses

Mediastinal masses are classified according to their location as anterior, middle and posterior mediastinal masses; common ones are summarized in Table 8.

Thymic Masses

Normal thymus (Fig. S16) should not be mistaken for an anterior mediastinal mass or right upper lobe collapse. Sail sign, cardiothymic notch and wave sign of Mulvey are described in normal thymus. Hyperplasia of the thymus is the most common thymic lesion in children. Thymomas are rare in children. In younger children, the thymus can have a quadrilateral shape with convex margins, while in older children, it becomes more triangular [36].

Germ Cell Tumors

Teratomas (Fig. 9) are the most common germ cell tumors, and are seen as mutilocular cystic tumors with areas of fat (-100 HU), fluid (-10 to +10 HU) and calcification.

Fig. 11 Differentiating neuroblastoma. a CXR shows large posterior mediastinal mass (*arrows*) (note negative silhouette sign for left cardiac margin). b Contrast CT Chest axial image demonstrates a large heterogeneous left paraspinal mass with foci of calcification (*arrow*) and intraspinal extension (*small double arrows*)

Lymphoma (Fig. S17)

They are the most common cause for anterior mediastinal mass in children [37]. Homogenous mildly enhancing enlarged discrete or conglomerate lymph nodal masses can be seen involving one or more mediastinal compartments.

Lymphatic Malformation

These are malformations of well-differentiated lymphatic channels. On CT, they appear smooth, lobulated trans-spatial masses of fluid density with enhancing septae, which insinuate around mediastinal structures (Fig. S18).

Bronchogenic Cyst (Fig. 10)

These are seen on CT as thin-walled single, unilocular cyst filled with air, serous fluid (fluid density of -20 to +10HU) or mucoid fluid (higher density). They can be mediastinal or peripheral.



 Table 9 Common chest
 Benign
 Lymphangioma

 wall masses in children
 Benign
 Lymphangioma

 Venous malformation
 Lipoma

 Osteochondroma
 Fibrous dysplasia

 Malignant
 PNET (Askins tumor)

 Osteosarcoma
 Rhabdomyosarcoma

Ganglion Cell Tumors

They arise from sympathetic chain ganglia and could be malignant (neuroblastoma), intermediate (ganglioneuroblastoma) and benign (ganglioneuroma) tumors. On CT, they are seen as mildly enhancing paraspinal soft tissue masses which may show intraspinal extension. MRI is better to assess the intraspinal component. Malignant tumors can show calcification, hemorrhage and necrosis (Fig. S19). They can spontaneously differentiate into less malignant forms (Fig. 11).

Nerve Sheath Tumors

Nerve sheath tumors (schwannomas and neurofibromas) are well-defined lobulated paraspinal masses which show moderate enhancement and may show intraspinal extension (Fig. S20). MRI is better suited to assess the intraspinal extension.

Neuroenteric Cyst

These are posterior mediastinal cystic lesions of foregut origin, associated with spinal or nervous system anomalies. MRI is better suited to assess the associated CNS anomalies (Fig. S21). CT will help in depicting the vertebral anomalies.

defined fat attenuation lesion (white arrow), deep to the muscles in the

Chest Wall Masses (Table 9)

anterior chest wall

Primitive Neuroectodermal Tumor of the Chest Wall- PNET (Askin's Tumor)

Ewing's sarcoma/PNET are small round cell tumors showing varying degree of neuroectodermal differentiation. On CT, heterogeneously enhancing extrapleural mass with adjacent rib destruction can be seen (Fig. S22). Lung metastasis is common.

Osteosarcoma

Matrix mineralization with disorganized ossification helps in diagnosing an osteosarcoma (Fig. 12).

Lipoma

Fat density (-100HU) on CT is diagnostic (Fig. 13).

Fig. 12 Osteosarcoma. Contrast CT Chest axial images in soft tissue (a) and bone window (b) demonstrate, a large ossified mass (*arrow*) arising from the chest wall with underlying rib destruction. Pleural dissemination of the tumor is seen as calcification of the pleura (*short arrows*) and pleural effusion (***)





Table 10 Proposed classification scheme for pediatric diffuse lung disease

- I. Disorders more prevalent in infancy
- A. Diffuse developmental disorders
 - 1. Acinar dysplasia
 - 2. Congenital alveolar dysplasia
- 3. Alveolar-capillary dysplasia with pulmonary vein misalignment
- B. Growth abnormalities
 - 1. Pulmonary hypoplasia
 - 2. Chronic neonatal lung disease
 - a. Prematurity-related chronic lung disease
 - (bronchopulmonary dysplasia)
 - b. Acquired chronic lung disease in term infants
 - 3. Structural pulmonary changes with chromosomal abnormalities a. Trisomy 21
 - b. Others
 - 4. Associated with congenital heart disease in chromosomally normal children
- C. Specific conditions of undefined etiology
 - 1. Pulmonary interstitial glycogenosis
- 2. Neuroendocrine cell hyperplasia of infancy
- D. Surfactant dysfunction mutations and related disorders 1. SPFTB genetic mutations—PAP and variant dominant
 - histologic pattern
 - 2. SPFTC genetic mutations—CPI dominant histologic pattern; also DIP and NSIP
 - 3. *ABCA3* genetic mutations—PAP variant dominant pattern; also CPI, DIP, NSIP
 - Others with histology consistent with surfactant dysfunction disorder without a yet recognized genetic disorder

- II. Disorders not specific to infancy
- A. Disorders of the normal host
 - 1. Infectious and post infectious processes
 - Disorders related to environmental agents: hypersensitivity pneumonia, toxic inhalation
 - 3. Aspiration syndromes
 - 4. Eosinophilic pneumonia
- B. Disorders related to systemic disease processes
 - 1. Immune-related disorders
 - 2. Storage disease
 - 3. Sarcoidosis
 - 4. Langerhans cell histiocytosis
 - 5. Malignant infiltrates
- C. Disorders of the immunocompromised host
 - 1. Opportunistic infection
 - 2. Disorders related to therapeutic intervention
 - 3. Disorders related to transplantation and rejection syndromes
- 4. Diffuse alveolar damage of unknown etiology
- D. Disorders masquerading as interstitial disease
 - 1. Arterial hypertensive vasculopathy
 - 2. Congestive vasculopathy, including veno-occlusive disease
 - 3. Lymphatic disorders
 - 4. Congestive changes related to cardiac dysfunction
- III. Unclassified—includes end-stage disease, nondiagnostic biopsies, and those with inadequate material

CPI Chronic pneumonitis of infancy; DIP Desquamative cell interstitial pneumonia; NSIP Nonspecific interstitial pneumonia; PAP Pulmonary alveolar proteinosis

Cough and Breathlessness or Suspected Diffuse Parenchymal or Interstitial Lung Disease (Table 10)

Unlike in adults, most ILDs in children have an underlying cause. Knowledge about the clinical symptoms is essential for radiological interpretation. Based on the American Thoracic Society guideline, a classification for Childhood interstitial lung disease [38] has been proposed.

Disorders More Prevalent in Infancy

- A. Diffuse Developmental Disorders There is no definite role for imaging, as most affected will die in first few days of life.
- B. Growth Abnormalities It is due to insult to lung parenchyma in prenatal or post-natal period.

Fig. 14 Alveolar proteinosis. A 12-y-old boy with recurrent LRI. Chest radiograph (a) and axial CT thorax in lung window (b) with a zoomed portion (c), show diffuse ground glass opacities with intra and inter lobualar septal thickening – crazy paving appearance



Table 11 Differentials for crazy paving appearance on HRCT

Acute	Subacute/Chronic
Pulmonary edema Pulmonary infection (pneumocystis, bacterial, viral) Pulmonary hemorrhage Acute interstitial pneumonia Eosinophilic pneumonia	Alveolar proteinosis Interstitial pneumonia – NSIP, UIP with diffuse alveolar damage Organising pneumonia Lymphangitic spread of tumor

NSIP Nonspecific interstitial pneumonia; UIP Usual interstitial pneumonia

- **Pulmonary hypoplasia** (Fig. S23) Underdeveloped bronchi and alveoli [39].
- Chronic neonatal lung disease Bronchopulmonary dysplasia (BPD) (Fig. S24) occurs in preterm infants who require mechanical ventilation and / or oxygen therapy. In severe stage of BPD, bubbly lucencies can be seen in imaging [40, 41].

C. Specific Conditions of Undefined Etiology

• Neuroendocrine cell hyperplasia of infancy (NEHI) or Persistent tachypnea of infancy.

Typical HRCT features are geographic ground glass opacity centrally, especially in lingula and right middle lobe without other interstitial abnormalities [42].

• Pulmonary interstitial glycogenosis (PIG) or Infantile cellular interstitial pneumonitis (histiocytoid pneumonia).

It is frequently associated with growth disorders [42].

D. Surfactant Dysfunction Mutations and Related Disorders

Alveolar proteinosis – The chest radiograph and diffuse ground glass opacities and interlobular septal thickening giving the "crazy paving" appearance (Fig. 14). Crazy paving appearance is not specific for this condition and can be seen in other conditions [43] as mentioned in Table 11.

Disorders Not Specific to Infancy

- A. Disorders of the Normal Host
 - Infectious and post infectious process. Swyer James syndrome or MacLeod syndrome - result of post infectious obliterative bronchiolitis secondary to viral respiratory infection in infancy or childhood (Fig. S10).
 - **Eosinophilic pneumonia.** They are rare in pediatric population. Simple and chronic eosinophilic pneumonia can occur in children. Radiological findings (Fig. S25) need to be correlated with the presence of peripheral and pulmonary eosinophila to confirm the diagnosis [44].

B. Disorders Related to Systemic Disease Processes

• Cystic fibrosis. CT findings include multilobar bronchiectasis, bronchial wall thickening, mucoid impaction in dilated bronchi and mosaic perfusion (Fig. 15).

Fig. 15 Cystic fibrosis- HRCT sections of the chest in lung window (**a**, **c**, **d**) show multilobar moderate cylindrical bronchiectasis (few shown by *arrows*), diffuse bronchial wall thickening, mucoid impaction. Atrophy of the pancreas (*thick arrow*) is seen on the abdomen image (**b**)



 Table 12
 Differentials for lung cysts in children

Pathology	Characteristic imaging feature(s)
Lymphangioleiomyomatosis (LAM)	Evenly distributed small cysts with thin walls involve lung bases. No nodules
Emphysema – no nodules	Air space opacity with imperceptible wall- could be centriacinar, para septal or pan acinar. No nodules
Cystic bronchiectasis	Follow the course of the bronchial tree and are contiguous with one another
Sarcoidosis	Cysts are predominantly apical in location, perilymphatic nodules, adenopathy
Interstitial pneumonia	Subpleural cysts, honeycomb like appearance, architectural distortion. Diffuse ground-glass opacities in active form
Septic emboli	Thick walled cysts in a patient with sepsis
Pneumatoceles	Occurs in areas of previous consolidation

CT scoring systems can be used in assessing disease status and in follow up [45].

- Immune related disorders. Various connective tissue disorders can show lung involvement in the form of interstitial pneumonitis (Fig. S26), follicular bronchiolitis (Fig. S27), pleural effusions *etc.*
- Storage disorders

Niemann Pick disease (Fig. S28). Ground glass opacities and interstitial thickening ('Crazy paving') can be seen due to accumulation of lipidstoring foamy histiocytes in interlobular septa or alveolar spaces [46]. Presence of hepatosplenomegaly should raise the suspicion of a storage disease.

• Langerhans cell histiocytosis. Multiple bilateral small nodules with cysts of varying wall thickness are the typical features (Fig. S29). Involvement of other systems like liver, spleen, bones, lymph nodes, CNS should be looked for.

Differentials for lung cysts in children are given in Table 12.

- C. **Disorders of the Immunocompromised Host** Opportunistic infections of fungal, viral and bacterial etiologies can infect the lung (Fig. S30).
- D. Disorders Masquerading as Interstitial Disease
 - Idiopathic pulmonary arterial hypertension (PAH) (Fig. S31).

Miscellaneous

Congenital Diaphragmatic Hernia (Figs. S32 and 16)

There are 3 basic types of congenital diaphragmatic hernias (Table 13). Children with congenital diaphragmatic hernia can have variable degree of pulmonary hypoplasia.



Fig. 16 Bilateral Bochdalek hernia. A 45-d-old baby with breathing difficulty since birth. Frontal radiograph of the chest (**a**) shows herniation of bowel loops into the right hemi thorax with displacement of the mediastinum (*thick arrow*). Left retrocardiac opacity is also seen (*thin arrows*).

CT axial (**b**) coronal (**c**) and sagittal (**d**, **e**) images showing bilateral Bochdalek hernias with herniation of bowel loops (*curved arrow*) and kidney on right side (*double arrows*) and herniation of spleen (*single arrow*) and part of fundus of stomach on left

Table 13Diaphragmatic hernias

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Туре	Location	Common contents	Diaphragmatic defect
Morgagni hernia (Fig. S32)	Anterior, more common on right	Can contain liver, omentum or bowel.	Near xiphoid, between the sternal and costal attachments of the diaphragm
Bochdalek hernia (Fig. 16)	Posterolateral, common on the left side	Large, usually contains bowel	Through lumbocostal triangle or Bochdalek's foramen formed by the incomplete closure of the pericardioperitoneal canals by the pleuroperitoneal membrane
Hiatus hernia	Through the esophageal hiatus	Stomach	

Conclusions

Computed tomography, when performed optimally for select conditions, can be a 'one-stop-shop' and problem solving imaging modality. To reduce radiation to the individual, and thus the community, due importance should be given by the entire team towards justification (selection of cases) and optimization of the study.

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