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Mulitidetector Computedtomography in the Evaluvation of Childhood Intestitial Lung Diseases

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Abstract: *Childhood interstitial lung diseases (chILD) is a group of rare chronic and complex disorders of variable pathology. Diagnosis is challenging because of different imaging pattern and rarity of its occurrence . CT thorax of 15 patients, 24 days to 3 years, with symptoms of interstitial lung disease , were studied over a period of three years . Ground-glass opacities either isolated or associated with other findings like centrilobular nodules, air trapping, septal thickening, crazy-paving, consolidation was the predominant finding seen in CT. Certain chILD can be diagnosed with CT without the need of lung biopsy.*
Abreviations: *chILD = Childhood interstitial lung disease*

I. Introduction

Childhood interstitial lung disease (chILD) is a heterogeneous group of rare diffuse lung diseases that can result in considerable morbidity and mortality. Interstitial lung disease may affect not only the interstitium but also the alveoli, airways, blood vessels, lymphatic channels, and pleural spaces (1). Typical features of ILD include dyspnoea, diffuse infiltrates on chest radiographs, and abnormal pulmonary function tests with restrictive ventilatory pattern and/or impaired gas exchange. While there is some overlap with adult disease, ILD in children often has a very different clinical picture. The problem may occur immediately after birth or may present at any age in childhood. Generally, the causes of these diseases are unknown but they may be inherited. Genetic testing, echocardiography, Computed tomography , bronchoscopy , lung biopsy are required in order to establish a precise diagnosis .Diagnosis is usually missed due to lack of clinical suspicion of the disease.

This study is done to evaluate various imaging patterns in childhood interstitial lung diseases.

II. Materials and Methods

The study included CT thorax of 15 interstitial lung disease patients of age group of 24 days to 3 years. Common symptoms were unexplained respiratory distress in a newborn, fast breathing, failure to thrive, typically dry cough and wheeze in the absence of respiratory tract infection. The duration of the study was three years .

Table 1 Ct Scan Protocols and Reconstructions

Scan range	Lung apex to diaphragm
Respiratory gating	No
Contrast media	No
Scanning technique	6 helical scanning
Matrix	512x512
Field of view	32 cm
Slice thickness	0.625 mm
Slice thickness of reformatted images	1.25 mm

Each CT was studied independently by 2 radiologists, each with10 years of experience in paediatric imaging. Pattern observed in each case was recorded. Presence and extent of geographic ground-glass opacities, mosaic attenuation , air trapping with decreased vascularity ,linear and reticular opacities, interstitial thickening, patchy consolidation, bilateral pleural effusion, crazy-paving, poorly defined centrilobular nodules, cysts, honey combing, traction bronchiectasis were recorded. (tab.2)

III. Results

Table 2 Patient Characteristics and Radiological Features

S.NO	Age of the patient	Gender	Clinical features	CT imaging findings	diagnosis
1	3 years	M	Cough, wheeze , breathlessness	mosaic perfusion and air trapping with decreased vascularity and central bronchiectasis	Bronchiolitis obliterans
2	2 years 5 months	M	Tachypnea and hypoxia	air trapping in a mosaic attenuation pattern with diffuse geographic ground-glass opacities esp. in right middle lobe, lingula	Neuroendocrine cell hyperplasia of infancy
3	10 days	M	Premature with respiratory distress	hyper lucent areas, linear and reticular opacities	Bronco pulmonary dysplasia
4	2 years	F	Cough and breathlessness	hyper lucent lung, mosaic attenuation and air trapping with decreased vascularity	Bronchiolitis obliterans
5	24 days	F	Premature with respiratory distress	air trapping, reticular and linear areas of opacity, sub segmental atelectasis	Bronco pulmonary dysplasia
6	2 years	F	Respiratory distress, hypoxia	ground glass opacities	Desquamative interstitial pneumonitis
7	3 years	M	Respiratory distress	multiple irregular cysts	pulmonary Langerhans cell histiocytosis (LCH)
8	1 year 4 months	M	Tachypnea	geographic ground-glass opacities in bil lower lobes, right middle lobe and lingula	Neuroendocrine cell hyperplasia of infancy
9	2years 2 months	M	Rhino rhea and breathlessness	mosaic attenuation and air trapping, central bronchiectasis	Bronchiolitis obliterans
10	2 years	F	Cough, breathlessness,	ground-glass opacity surrounded by consolidation	organising pneumonia
11	1 year	M	Tachypnea, failure to thrive	consolidation with fibrosis, ground glass opacities, nodules, peripheral air trapping	Neuroendocrine cell hyperplasia of infancy
12	3 years	M	dry cough and wheeze in the absence of respiratory tract infection	Patchy consolidation, air trapping , interstitial thickening, ground glass opacity, bilateral pleural effusion	Bronchiolitis obliterans organising pneumonia
13	20 days	M	Respiratory distress	Ground-glass opacities mixed with hyper inflated or hyper lucent areas.	Pulmonary interstitial glycogenosis
14	28 days	F	fast breathing, failure to thrive	ground-glass opacities with septal thickening, crazy-paving, consolidation	Pulmonary alveolar proteinosis
15	2 years	M	Respiratory distress	poorly defined centrilobular nodules, ground-glass opacities, air trapping	Hypersensitivity pneumonitis

Table- 3 Ct Imaging Pattern Analysis

S.no	Radiological pattern	No of patients	Percentage %	Diagnosis
1	**Ground-glass opacities**	6	40	
	Isolated ground glass opacities	1	6	Desquamative interstitial pneumonitis
	Diffuse geographic ground-glass opacities in right middle lobe, lingula ± other lobes with air trapping in lower lobes	3	20	Neuroendocrine cell hyperplasia of infancy
	ground-glass opacities with poorly defined centrilobular nodules, air trapping	1	6	Hypersensitivity pneumonitis
	ground-glass opacities with septal thickening, crazy-paving,	1	6	Pulmonary alveolar proteinosis
2	**Consolidation**	2	13	
	Patchy consolidation	1	6	Bronchiolitis obliterans organising pneumonia
	ground-glass opacity surrounded by consolidation	1	6	organising pneumonia
3	Nodules	1	6	Hypersensitivity pneumonitis
4	Mosaic attenuation(ground glass opacities with air trapping) ± pulmonary vascular attenuation ,bronchial wall thickening	3	20	Bronchiolitis obliterans
5	**Linear/reticular opacities**	3	20	Bronco pulmonary dysplasia, Bronchiolitis obliterans organising pneumonia
6	**Cysts**	1	6	Pulmonary Langerhans cell histiocytosis

IV. Discussion

CHILD syndrome requires the presence of at least 3 of the following 4 criteria in the absence of other known disorders: (a) respiratory symptoms (cough, rapid breathing, or exercise intolerance), (b) signs (resting tachypnea, crackles, retractions, digital clubbing, failure to thrive, or respiratory failure), (c) hypoxemia, and (d) diffuse chest in filtrates on chest X-ray or computed tomography (CT) scan. (2)

Pulmonary abnormalities found on HRCT in our study predominantly consists of ground-glass opacities(40%), consolidation(13%), nodules(6%), mosaic attenuation(20%), linear/reticular opacities(20%), or cysts(6%).

Ground-glass opacity is defined as hazy increased attenuation of the lung with preserved visibility of the bronchovascular structures. Ground-glass opacity is very nonspecific and can be seen in lung with increased capillary blood volume, such as from shunting, or in lung affected by processes that partially remove or replace the air in the alveoli with cells, fluid, or other material.

Ground-glass opacity (40%) is seen in Desquamative interstitial pneumonitis, Neuroendocrine cell hyperplasia of infancy, Hypersensitivity pneumonitis and Pulmonary alveolar proteinosis. Other conditions with ground glass opacities are Pulmonary interstitial glycogenosis, inborn errors of surfactant metabolism(3), Pulmonary lymphangiectasia, lymphangiomatosis, pulmonary veno-occlusive disease, Pulmonary alveolar microlithiasis(1), Pulmonary Langerhans cell histiocytosis(4). Ground-glass opacities can also be observed physiologically in underinflated lungs, particularly in the dependent lung bases of infants on images acquired at expiration or shallow inspiration(1).

consolidation is the airspace opacification that obscures the pulmonary vessels. consolidation(13%) is seen in Bronchiolitis obliterans organising pneumonia and organising pneumonia. Consolidation can also be seen eosinophilic pneumonia, aspiration pneumonia, acute interstitial pneumonia (AIP), pulmonary haemorrhage syndromes, and alveolar pulmonary oedema.(1)

Pulmonary nodules can be classified according to their distribution as perilymphatic, miliary, or centrilobular. Perilymphatic nodules are found along the interlobular septa, interlobar fissures, and bronchovascular bundles. Nodules are seen in Hypersensitivity pneumonitis (6%).

Miliary nodules have a random distribution and may be seen in Langerhans cell histiocytosis (LCH) . Centrilobular nodules are found within the secondary pulmonary lobule and are separated from the pleural fissures and interlobular septa by a distance of several millimetres (1) . Centrilobular nodules with ground-glass attenuation are commonly seen in hypersensitivity pneumonitis(4), Bronchiolitis, cystic fibrosis(5), asthma, immotile cilia syndrome(5), idiopathic pulmonary hemosiderosis (IPH), and pulmonary veno-occlusive disease(PVOD). The tree-in-bud pattern refers to branching centrilobular opacities that resemble a budding tree, and is most commonly observed in disorders associated with endobronchiolar plugging, such as cystic fibrosis(5) and Bronchiolitis , but can also be seen in diseases such as capillaritis that affect the intralobular vessels(1)

Mosaic attenuation is defined as heterogeneous areas of differing lung attenuation . This heterogeneous pattern of attenuation is the result of diverse causes that include diseases of the small airways, pulmonary vasculature, alveoli, and interstitium, alone or in combination. Small airways disease can be a primary disorder, such as respiratory bronchiolitis or constrictive bronchiolitis, or be part of parenchymal lung disease, such as hypersensitivity pneumonitis, or large airways disease, such as bronchiectasis and asthma. Vascular causes resulting in mosaic attenuation are typically chronic thromboembolic pulmonary hypertension, which is characterized by organizing thrombi in the elastic pulmonary arteries, or pulmonary arterial hypertension, a heterogeneous group of diseases affecting the distal pulmonary arterioles(6). Mosaic attenuation (20%) observed in Bronchiolitis obliterans and Neuroendocrine cell hyperplasia of infancy.

A cyst appears on CT as a round low-attenuating structure with a well-defined interface with normal lung. Cysts usually contain air but occasionally contain fluid or solid material. Cyst is a rare finding predominantly noted in LCH(5%). other conditions with thin-walled cysts are LIP, NSIP, lung growth disorders, disorders of surfactant metabolism, and pulmonary alveolar microlithiasis . Thicker-walled cysts seen in honeycombing reflect the dissolution of alveoli and loss of acinar architecture associated with pulmonary fibrosis(1).

V. Conclusion

Ground-glass opacities either isolated or associated with other findings like centrilobular nodules, air trapping, septal thickening, crazy-paving, consolidation is the predominant finding seen in CT scan of childhood interstitial lung disease patients.. Multidetector CT may aid diagnosis,identify a site for biopsy, and help monitor the disease.

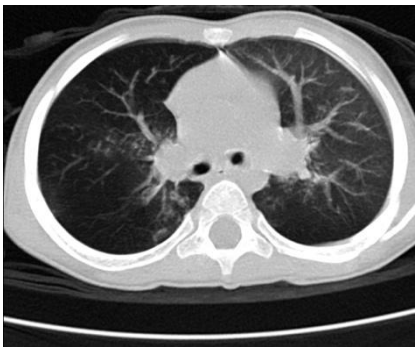

Fig1, HRCT reveals hyperlucent lung, mosaic attenuation, air trapping with decreased vascularity and bronchectasis in a case of bronchiolitis obliterans

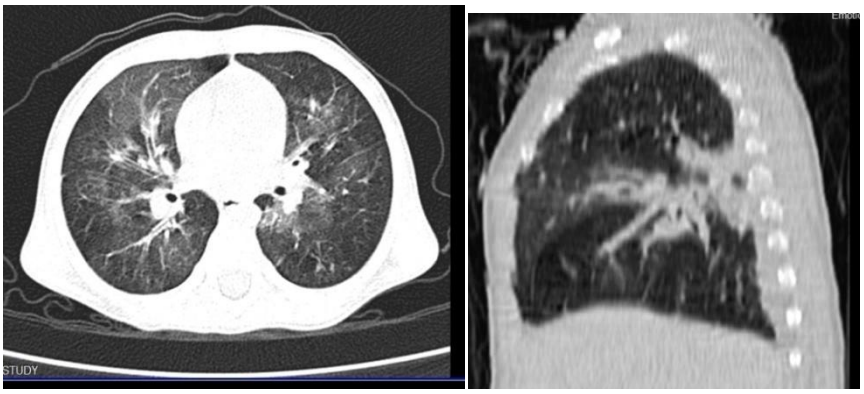

Fig 2&3, 2 year male child with neuroendocrine hyperplasia of infancy. CT axial and sagittal reformatted images shows air trapping in a mosaic attenuation pattern with geographic ground-glass opacities in right middle lobe , lingula (very characteristic of NEHI) and bilateral lower lobes

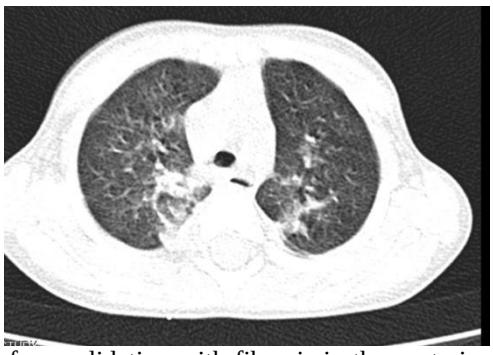

Fig 4, CT shows areas of consolidation with fibrosis in the posterior segment of bilateral upper lobe ,ground glass opacities ,nodules in the central part of bilateral lung with peripheral air trapping in a 1 year male child suggestive of interstitial lung disease.

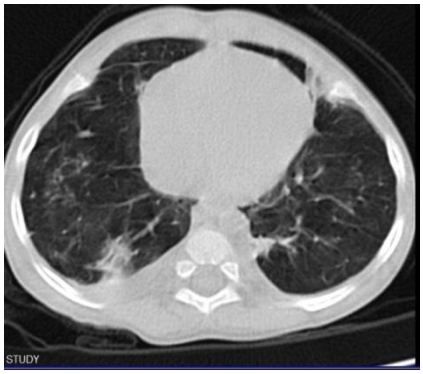

Fig 5,Areas of peripheral patchy consolidation, multi focal air trapping , interstitial thickening, areas of ground glass opacity,mild bilateral pleural effusion (R > L) seen in bronchchiolitis obliterans organising pneumonia.

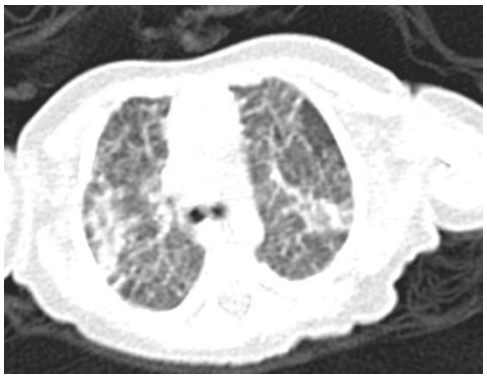

Fig 6, 24 days female child with bronchopulmonary dysplasia. CT shows hyperlucent areas, linear and reticular opacities.

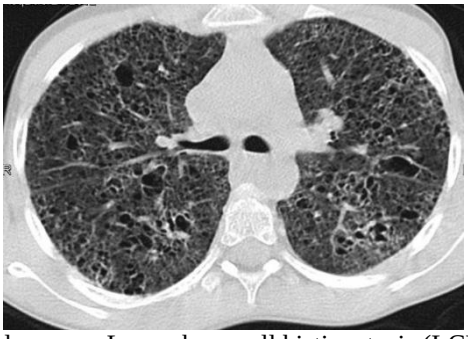

Fig 7, 3 years male child with pulmonary Langerhans cell histiocytosis (LCH). CT shows multiple irregular cysts in bilateral lung fields.

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