

Role of MSCT in Diagnosis of Interstitial Lung Disease in Children

Essay

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قالوا

سبحانك لا علم لنا
إلا ما علمتنا إنك أنت
العليم الحكيم

صدق الله العظيم

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List of Abbreviations

Abb.	Full term
<i>3-D</i>	<i>Three-dimensional</i>
<i>ABPA</i>	<i>Allergic bronchopulmonary aspergillosis</i>
<i>AIP</i>	<i>Acute interstitial pneumonia</i>
<i>ALARA</i>	<i>As low as reasonably achievable</i>
<i>ARDS</i>	<i>Acute respiratory distress syndrome</i>
<i>BG</i>	<i>broncho-centric granulomatosis</i>
<i>CEP</i>	<i>Chronic eosinophilic pneumonia</i>
<i>CFA</i>	<i>Cryptogenic fibrosing alveolitis</i>
<i>chILD</i>	<i>Children's interstitial lung disease</i>
<i>CT</i>	<i>Computed tomography</i>
<i>CTA</i>	<i>CT angiography</i>
<i>CVL</i>	<i>Central venous lines</i>
<i>CXR</i>	<i>Chest radiographs</i>
<i>DAD</i>	<i>Diffuse alveolar damage</i>
<i>EMT</i>	<i>Epithelial-mesenchymal transition</i>
<i>GM-CSF</i>	<i>Granulocyte macrophage colony-stimulating factor</i>
<i>HVL</i>	<i>Half value layer</i>
<i>IHS</i>	<i>Idiopathic hypereosinophilic syndrome</i>
<i>ILDs</i>	<i>Interstitial lung diseases</i>
<i>IPF</i>	<i>Idiopathic pulmonary fibrosis</i>
<i>IPH</i>	<i>Idiopathic pulmonary hemosiderosis</i>
<i>LCH</i>	<i>Langerhans cell histiocytosis</i>
<i>LIP</i>	<i>Lymphocytic interstitial pneumonitis</i>
<i>MPR</i>	<i>Multi-planar reconstruction</i>

List of Abbreviations Cont...

Abb.	Full term
<i>MSCT</i>	<i>Multislice CT</i>
<i>NSIP</i>	<i>Nonspecific interstitial pneumonia</i>
<i>PAP</i>	<i>Pulmonary alveolar proteinosis</i>
<i>PCH</i>	<i>Pulmonary capillary hemangiomatosis</i>
<i>PFTs</i>	<i>Pulmonary function tests</i>
<i>PICC</i>	<i>Peripherally inserted central catheter</i>
<i>PIE</i>	<i>Pulmonary infiltrate with eosinophilia</i>
<i>Psi</i>	<i>Pounds per square inch</i>
<i>PVOD</i>	<i>Pulmonary veno-occlusive disease</i>
<i>SDMs</i>	<i>Surfactant-dysfunction mutations</i>
<i>SPE</i>	<i>Simple pulmonary eosinophilia</i>
<i>TGF- β</i>	<i>Transforming-growth factor-β</i>

INTRODUCTION

Interstitial lung diseases (ILDs) in childhood are a diverse group of conditions that primarily involve the alveoli and peri-alveolar tissues, leading to derangement of gas exchange, restrictive lung physiology, and diffuse infiltrates on radiographs. Because ILDs can involve the distal airspaces as well as the interstitium, the term diffuse infiltrative lung disease has been suggested. This nomenclature may be more accurate than ILD, but children's interstitial lung disease (chILD) has become the preferred term (*Dishop, 2011*).

Many types of chILD following some type of injury to the distal airspaces, such as adenoviral infection or exposure to organic dust, resulting in damage to the epithelial or endothelial layers and the associated basement membrane. Inflammation is present in many types of chILD, and many forms of chILD are triggered by inflammatory events, such as infection or hypersensitivity. Almost every type of inflammatory cell, including eosinophils and mast cells, have been described in various types of chILD and can interact with fibroblasts and other parenchymal cells. However, lung inflammation does not necessarily result in fibrotic remodeling, and fibrosis can occur in the absence of inflammation; therefore, inflammation has a prominent, but not a central, role in lung remodeling and fibrosis (*Das et al., 2011; Vece et al., 2011*).

Resolution of fibrotic remodeling involves a complex series of orderly steps, including matrix breakdown and restructuring, re-epithelialization, and apoptosis of fibroblasts and inflammatory cells. Fibrotic remodeling is responsible for most of the morbidity and mortality associated with chILD. Remodeling of distal airspaces resulting hypoxemia. Persistent hypoxemia results in pulmonary hypertension and vascular remodeling, leading to cor pulmonale, the increased work of breathing associated with reduced compliance results in increased energy expenditure, which, combined with the effects of inflammatory mediators, can result in cachexia. Portions of the lung may be replaced by fibrotic septae between dilated airspaces, the so-called honeycomb changes of end stage interstitial disease. Although the events described above are necessary for repair of the injured lung, excessive activation or failure of resolution of any of these pathways can result in disabling fibrosis (*He et al., 2014*).

As a result of the rarity of ILDs in children and the important differences between childhood ILD and ILDs that affect adults, a great deal of confusion surrounds their nomenclature, classification, and management. Idiopathic pulmonary fibrosis (IPF, also known as cryptogenic fibrosing alveolitis [CFA], the most prominent adult ILD, mostly occurs after the fifth decade of life; this entity is not found in children. Unlike in adults, most ILDs in children are found to have an underlying cause. In addition, the clinical significance of the