What the Pulmonologist Needs From the Radiologist

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Childhood interstitial lung disease (chILD) is a subset of pediatric diffuse lung disease.

- Rare, heterogenous group of lung diseases that affect the airways, alveolar spaces and interstitium.
- Characterized by:
  - Diffuse abnormalities on imaging
  - Impaired gas exchange
- Childhood interstitial lung disease (chILD)
  - Occurs in a variety of clinical contexts including isolated pulmonary disorders, as a consequence of environmental exposures, in the setting of systemic disorders and in the setting of unknown etiology.
- Though some forms are similar, most childhood ILD is markedly different from adult ILD in etiology, treatment and outcome.

Childhood ILD includes a distinct set of diseases only seen in infants.

<table>
<thead>
<tr>
<th>Category</th>
<th>Specific Diagnosis</th>
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<tbody>
<tr>
<td>Diffuse Developmental Disorders</td>
<td>Acinar dysplasia, Congenital alveolar dysplasia, Alveolar-capillary dysplasia with pulmonary vein misalignment</td>
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<tr>
<td>Lung Growth Abnormalities</td>
<td>Pulmonary hypoplasia, Chronic neonatal lung disease (BPD), Associated chromosomal disorders (trisomy 21, others), Associated with congenital heart disease</td>
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<tr>
<td>Specific conditions of uncertain cause</td>
<td>Neuroendocrine cell hyperplasia of infancy (NEHI), Pulmonary interstitial glycogenosis (PIG)</td>
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<td>Surfactant Dysfunction</td>
<td>SFTPB, SFTPC, ABCA3, NKX2.1/TTF1, Histology consistent with surfactant dysfunction but without recognized genetic cause</td>
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The diagnosis of “chILD syndrome” is made when 3 of the following 4 criteria are met.

1. **Respiratory Symptoms**
   - Cough, increased work of breathing at baseline, exercise intolerance

2. **Respiratory Signs**
   - Tachypnea at rest, digital clubbing, crackles, failure to thrive, respiratory failure

3. **Hypoxemia**

4. **Diffuse parenchymal abnormalities on imaging**

Diagnosis of the specific chILD diagnosis allows for optimization of therapies and resources for the patient and their family.

- Systemic steroids
- Steroid sparing therapies
- Chemotherapy/Immunotherapy
- Genetic counseling
- Supportive Care
  - Oxygen
  - Ventilatory support
  - Nutrition
  - Immunizations
  - Control of comorbidities
An individualized stepwise approach is recommended for evaluation of ILD in a child.

- Exclude more common diseases causing similar signs and symptoms
  - Cystic fibrosis, primary ciliary dyskinesia, chronic aspiration, immunodeficiency, cardiac disease
- Computed tomography
  - Inspiratory and expiratory views are essential
- Bronchoscopy with bronchoalveolar lavage
- Genetic testing
- Lung biopsy
Though lung biopsy remains the gold standard for diagnosis of chILD, specific forms of ILD can be diagnosed or suggested without lung biopsy.

- Genetic testing for disorders of surfactant dysfunction
  - SFTPB, SFTPC, ABCA3, NKX2.1/TTF1, GMCSF receptors α and β
- CT findings can be suggestive of certain diseases
  - Surfactant dysfunction disorders
  - Bronchiolitis Obliterans
- CT findings can be specific for certain disorders
  - Neuroendocrine cell hyperplasia of infancy (NEHI)

Brody AS, Guillerman RP et al. Neuroendocrine Cell Hyperplasia of Infancy: Diagnosis with High Resolution CT. Am J Roentgenol 2010; 194:238–244
Computed tomography can define the presence, extent and pattern of disease in a child with ILD and may avoid lung biopsy.

- ILD can be suggested based on the distribution, size and/or quantity of the following findings on CT interpretation:
  - Hyperinflation; vascular attenuation
  - Ground glass opacities
  - Linear, reticular, or nodular densities
  - Peribronchial or septal thickening
  - Bronchiectasis
  - Cystic lesions
  - Other: calcifications, adenopathy, pleural thickening, and pleural fluid

- CT chest can also help determine optimal area for lung biopsy when needed.

A set of identical twins with NEHI.

Ground glass opacities in the right middle lobe, lingula, and perihilar regions, without other abnormalities, demonstrates a pattern consistent with NEHI.

Toddler with Post Infectious Bronchiolitis Obliterans

Presented to pulmonary 1-2 months s/p RSV bronchiolitis infection with tachypnea, retractions, wheeze, hypoxemia and FTT. Chest CT confirmed diagnosis of Bronchiolitis Obliterans Syndrome (BOS).
1 year old child testing positive for SFTPC mutation.

Chest CT of a 1 year old born at term, requiring O2 in the first week of birth with a history of multiple hospitalizations for hypoxemia, tachypnea and baseline respiratory distress in the setting of a family history of pulmonary fibrosis. Genetic sequencing confirmed SFTPC mutation (L181V).
Safe and effective imaging in the evaluation of a child for ILD should include multidisciplinary communication.

- Imaging of infants in the evaluation of ILD may require sedation and controlled ventilation due to their clinical symptoms and the clarity of the images needed.
  - Extreme Tachypnea
  - Need to determine presence of hyperinflation, air trapping, concerns for true abnormalities versus atelectasis/artifact
- Communication between radiologist, anesthesiologist and pulmonologist prior to imaging allows for best imaging outcomes.
References and Suggested Reading

• Brody AS, Guillerman RP et al. Neuroendocrine Cell Hyperplasia of Infancy: Diagnosis with High Resolution CT. Am J Roentgenol 2010; 194:238–244
• Guillerman RP. Imaging of Childhood Interstitial Lung Disease. Pediatric Allergy, Immunology, and Pulmonology; 2010; 23:1: 43-68