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Pediatric Interstitial Lung Disease-Approach to Diagnosis and Management-Review

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Abstract

Interstitial lung disease (ILD) consists of a diverse group of disorders that involve the pulmonary parenchyma and interfere with gas exchange. "Diffuse parenchyma lung disease" or "diffuse lung disease". ILD is rare in childhood.

- *Children with ILD may present with respiratory failure, or with more indolent or chronic symptoms including tachypnea, hypoxemia, retractions, cough, exercise intolerance, failure to thrive, and gastro esophageal reflux.*

- *ILD should be considered in any neonate who presents with unexplained respiratory failure, or in infants and children with a normal birth history who present with persistent tachypnea, crackles, hypoxemia, chronic cough, or clubbing of the digits.ILD should also be considered in late preterm or preterm infants who present with chronic lung disease out of proportion to the degree of prematurity or other known co morbidities*

- *Diagnostic studies in children with suspected interstitial lung disease is to exclude more common causes of chronic respiratory symptoms: Cough, wheeze and dyspnoea Chest deformity, stunting and failure to thrive. clubbing, halitosis sputum production, cyanosis, or cor-pulmonale. Chest X-Ray finding mainly Interstitial infiltrate (Discrete, Linear, Nodular or reticulonodular shadows < 2mm) in diffuse distribution in both lung. In advanced stage fibrosis is extensive & lungs are shrunken & reduced in volume. normal CXR doesn't exclude ILD.* ***Spirometry*** *–In ILD there is restrictive defect, tidal volumes – small, vital capacity / TLC – Reduced.* ***HRCT of chest****: Ground glass opacity, Reticulonodular Shadowing Honey comb lung (small, uniform sized, cystic spaces representing patent bronchioles). It provides quantitative assessment of pulmonary fibrosis.*

Keywords: Interstitial, Lung, Disesases, Children, Corticosteroid

Introduction

Interstitial lung disease consists of a diverse group of disorders that involve the pulmonary parenchyma and interfere with gas exchange. "Diffuse parenchyma lung disease" or "diffuse lung disease" ILD is rare in childhood. Children with ILD may present with respiratory failure, or with more indolent or chronic symptoms including tachypnea, hypoxemia, retractions, cough, exercise intolerance, failure to thrive, and gastro esophageal reflux. [1, 2].

ILD should be considered in any neonate who presents with unexplained respiratory failure, or in infants and children with a normal birth history who present with persistent tachypnea, crackles, hypoxemia, chronic cough, or clubbing of the digits. ILD should also be considered in late preterm or preterm infants who present with chronic lung disease out of proportion to the degree of prematurity or other known co morbidities [1-3].

Methods

Data Extraction

By performing a searched in Pub Med and the Cochrane Library and EMBASE with the keywords: (ILD * Pediatric) AND (diagnostic approach and treatment of corticosteroids limited for clinical trials or systematic reviews of ILD. Relevant study areas were identified, and, for each area, a literature search was carried out based on a predefined series of key clinical questions. and the strategies included filters to limit the results by study type (reviews, randomized controlled trials and other types of experi-

mental research) to English language material [4-6]. The present manuscript was organized into three main sections: Definitions, diagnostic approach and Treatment of interstitial lung diseases.

Results

ILD should be considered in any neonate who presents with unexplained respiratory failure, or in infants and children with a normal birth history who present with persistent tachypnea, crackles, hypoxemia, chronic cough, or clubbing of the digits. ILD should also be considered in late preterm or preterm infants who present with chronic lung disease out of proportion to the degree of prematurity or other known co morbidities [1, 2].

Types of chILD?

Some types of chILD are genetic and passed through families. Some types are caused by an environmental or infectious trigger. Some have an unknown cause. More and more genetic causes for chILD are discovered as we learn more about genes. Examples of types of chILD: ■ Bronchiolitis Obliterans ■ Chronic Bronchiolitis ■ Connective tissue associated lung disease ■ Cryptogenic Organizing Pneumonia (COP) ■ Alveolar Capillary Dysplasia with misalignment of the pulmonary veings ■ Hypersensitivity Pneumonitis ■ Capillaritis ■ Neuroendocrine Hyperplasia of Infancy (NEHI), also known as Persistent Tachypnea of Infancy ■ Pulmonary Interstitial Glycogenosis (PIG) ■ Surfactant dysfunction mutation [4-7].

Diagnostic Approach

Diagnosis should be done on the basis of clinical history, physical examination and appropriate use of further investigations.

The purpose of history-taking and physical examination and relevant investigations are to confirm the diagnosis of ILD and also to exclude common problems like the preschool child with wheezing disorder, asthma and congenital heart diseases [6-9].

Symptoms

- Chronic cough, wheeze and dyspnoea
- Sputum production
- Chest deformity
- Halitosis
- Features of right heart failure
-
- **Examination**

1) Tachypnoea
2) Hypoxia
3) Widespread crackles
4) Cyanosis
5) Clubbing
6) Failure to thrive

- Diagnostic studies in children with suspected interstitial lung disease to assess extent and severity of disease:

1. Chest Radiographs
   Interstitial infiltrate (Discrete, Linear, Nodular or reticulonodular shadows < 2mm) in diffuse distribution in both lungs. In advanced stage fibrosis is extensive & lungs are shrunken & reduced in volume. Normal CXR doesn't exclude ILD (6-9)

2. Chest Computed Tomography Scan
   Ground glass opacity, Reticulonodular shadowing Honey comb lung (small, uniform sized, cystic spaces representing patent bronchioles). It provides quantitative assessment of pulmonary Interstitial infiltrate (Discrete, Linear, Nodular or reticulonodular shadows < 2mm) in diffuse distribution in both lungs. (6-9)

3. Electrocardiogram/ Echocardiogram

4. Pulmonary function Studies

a) To exclude chronic respiratory diseases like (**Cystic Fibrosis, Gastroesophageal Reflux Desiseas, Asthma, and Bronchiolitis Obliterans**) **[6-10]:**

5. Sweat chloride testing for diagnosis of Cystic fibrosis

6. Evaluation for gastro esophageal reflux and recurrent aspiration (such as barium swallow, pH/impedance probe and others)

- Bronchoscopy & Bronchoalveolar lavage – Bronchiolitis obliterans (Narrowing of terminal bronchiole)

a. Bronchiolitis obliterans is a histological diagnosis which is confirmed by biopsy and histopathology
b. Bronchoalveolar lavage is useful in excluding pulmonary eosinophilia, hypersensitivity pneumonitis.

- MT, sputum for AFB, gene x-pert and culture, stool for gene x-pert ultra: Evaluation of TB
- Echocardiogram – Evaluation for CHD.

7 Cultures and testing for infectious etiologies

B) To identify systemic disorders predisposing to ILD [4-6].

8. PID panel: Evaluation of Immunodeficiency/Immune dysfunction.
9. ANA, ANCA: Evaluation of connective tissue disorder like (Sarcoidosis, Auto- immune disease, auto immune vacuities)
10. Allergic Panel test/Hypersensitivity pneumonitis panel.
11. Genetic studies for surfactant dysfunction.

Treatment [11-14]

- Supportive therapy and pharmacologic interventions, tailored to the type of ILD:
- Criteria to start treatment-
- Presence of severe/worsening symptoms
- Younger age of onset
- Shorter duration of illness

Supportive Therapy

1. Limiting exposure to cigarette smoke and other inhaled irritants
2. Oxygen therapy for hypoxemia
3. Supervised exercise
4. Bronchodilators for reversible airway obstruction
5. Aggressive treatment of intercurrent infections
6. Standard childhood vaccinations
7. RSV Immunoprophylaxis

Specific Treatment [12-15]

8. Specific treatment is available for some ILD disorders

9. Antimicrobials for certain infections, management of swallowing dysfunction and/or reflux in patients with chronic aspiration, avoidance of the offending antigen in hypersensitivity pneumonitis, and whole lung lavage for older children with pulmonary alveolar proteinosis
10. Glucocorticoids are the mainstay of therapy for many children with ILD because inflammation and inappropriate cellular proliferation are thought to play an important role in pathogenesis of many ILD subtypes
11. Prednisolone given in a dose of 1 – 1.5 mg for 6 – 12 weeks followed by maintenance dose of 15 – 20mg daily for 1 – 2 years or longer
12. I.V. Cyclophosphamide given as intermittent pulse therapy (1 – 1.3 gm / month) along with pred. therapy provides symptomatic relief.
13. Combination of low dose prednisolone with Azathioprine – maintenance Tt. For 2-3 yrs.
14. Lung transplantation

Summary

Children's Interstitial and Diffuse Lung Disease (chILD) is a group of rare lung diseases found in infants, children and teens. There are many types of chILD. The different types of chILD and have some of the same symptoms. The symptoms, however, may vary in how severe they are. Diagnosis of ILD in children is challenging in developing country like Bangladesh because there are no available diagnostic facilities. Treatment is also long-term steroid with some cases need cyclophosphamide and azathioprine.

Ethics Approval and Consent to Participate

Since it is a review paper, the study did not need ethical committee approval.

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