EDITORIAL

Interstitial Lung Disease in Children

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Interstitial lung diseases (ILD) in children comprise a diverse group of rare conditions involving the interstitium as well as the distal airspaces that result in restrictive lung physiology and derangements in oxygenation. The expression and outcome of pediatric ILD differs from adult ILD as the disease occurs during the growing phase of the lung and extrapolation of information obtained from adult studies has been debated. Several classifications for ILD have been proposed but none have been found to be entirely satisfactory especially in children. Since ILD comprises of heterogenous group of disorders, it is logical to approach the disease as unknown etiology (idiopathic), known etiology and ILD unique to infancy.

Authors of the study published in this issue of Indian Pediatrics [1] have done extensive research and classified patients with ILD as definite and possible subgroups. Since ILD occurs frequently in the younger children and causes are different from adults, future research should concentrate on younger patients. Idiopathic ILD requires a tissue diagnosis. The lung histological patterns that can be observed in ILD have been reviewed by the ATS/ERS [2]. Some idiopathic disorders described in adults like desquamative interstitial pneumonia (DIP), non-specific interstitial pneumonia (NSIP), lymphocytic interstitial pneumonia (LIP) are reported in children also. NSIP has been reported with connective tissue and surfactant disorders and LIP with immunodeficiency states.

It is our observation that in children with idiopathic ILD the mean survival duration was about two and half years. Acute interstitial pneumonia (formerly Hamman-Rich syndrome), rapidly progresses to fatal respiratory failure was also reported in children [3]. Interstitial lung disease unique to infancy includes cellular intersititial pneumonitis of infants, chronic pneumonitis of infancy, familial DIP, idiopathic pulmonary fibrosis of infancy, infantile pulmonary hemosiderosis, surfactant dysfunction disorders and persistent tachypnea of infancy. [4]. The study published in this issue [1] though discusses both unknown and known causes, contributes significant information on the latter group. Chronic aspiration, pulmonary hemosiderosis, pulmonary alveolar proteinosis, Langerhans cell histiocytosis, hypersensitivity pneumonitis, sarcoidosis, and lymphocyte infiltrative disorders come under known etiology and majority of them may be diagnosed without biopsy where bronchoalveolar lavage (BAL) plays an important role [5].

Except the acute form, children with ILD have symptoms for years before the diagnosis. Due to rarity of the condition and lack of awareness, the victims are subjected to repeated chest skiagrams, antibiotics and anti-tuberculosis therapy. Cough, dyspnea, failure to thrive, exercise intolerance, retractions, inspiratory (velcro) crackles and hypoxemia are the features associated with initial presentation, and when these are not explained by an alternative diagnosis, ILD should be suspected. The usefulness of BAL and HRCT has been well emphasized in the study. HRCT should be included in the early diagnostic workup of ILD which may show geographical hyperlucency, septal thickening and ground glass opacity and to optimise spatial resolution thin slice cuts of one mm collimation is recommended [6].

In a resource poor setting, a systematic approach integrating strong suspicion, a thorough clinical evaluation and HRCT may be suffice for the diagnosis and biopsy is rarely indicated as the specimen obtained may not always be a representative sample [7] and this fact has been highlighted in the present study. In our opinion, lung biopsy can be included in the workup of "unknown etiology" group as it may throw more light on this medical enigma particularly in centers equipped with video assisted thoracoscopy. Although spirometry and pulse-oximetry do not provide specific information, they do play a role during follow-up. Authors of present study assigned ILD scores which may predict the outcome in individual cases.

Oral prednisolone or pulsed intravenous methyl prednisolone, singly or in combination with hydroxychloroquine, are commonly used drugs and children with significant disease are better treated with pulsed methyl prednisolone. The aim of treatment is to maintain the patient on the minimum dose of steroid compatible with clinical stability.

Collaborative multicentric studies in pediatric ILD focusing on molecular mechanisms, pathophysiology and natural history are the need of the hour which may save the affected children from an expensive diverse battery of investigations and inappropriate therapy. The research paper published in this issue will raise awareness about ILD among pediatricians.

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REFERENCES

1. Sankar J, Pillai MS, Sankar MJ, Lodha R, Kabra SK. Clinical profile of interstitial lung disease in children. Indian Pediatr;2013:50:127-33.
2. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus. Classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med. 2002;165:277-304.
3. Vijayasekaran D, Giridhar S, Gowrishankar NC, Nedunchelian K, Senguttuvan M. Pediatric interstitial lung disease. Indian Pediatr. 2006;43:899-903.
4. Dishop MK. Paediatric interstitial lung disease: classification and definitions. Paediatr Respir Rev. 2011;12:230-7.
5. de Blic J, Midulla F, Barbato A, Clement A, Dab I, Eber E, *et al*. Bronchoalveolar lavage in children. ERS Task Force on Bronchoalveolar Lavage in Children. European Respiratory Society. Eur Respir J. 2000;15:217-31.
6. Clement A. Task force on chronic interstitial lung disease in immunocompetent children. Eur Respir J. 2004;24:686-97.
7. Fan LL, Kozinetz CA, Deterding RR, Brugman SM. Evaluation of a diagnostic approach to pediatric interstitial lung disease. Pediatrics. 1998;101:82-5.