IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 24, Issue 2 Ser. 4 (February. 2025), PP 27-29
www.iosrjournals.org

Epidemiology, Clinical Presentation and Outcome of Children With Interstitial Lung Disease.

Dr. Murali Krishnan P[1], Dr. Anusha[2] Dr. Vijayasekar D[3]
(Associate Professor Department of Pediatrics, Sri Lakshmi Narayana Institute of Medical Sciences, BIHER University, India)
(Assistant Professor Department of Pediatrics Sri Lakshmi Narayana Institute of Medical Sciences, BIHER University, India)
(Professor, Institute of Child Health, Madras Medical College, The Tamil Nadu Dr. MGR Medical University, India)

Abstract
Background: *Pediatric interstitial lung disease presents (ILD) both diagnostic and therapeutic challenge. There are very few studies on interstitial lung disease in children because of its rarity and difficulty in diagnostic confirmation.*
Methods: *A retrospective study was done to analyze the epidemiological profile, clinical presentation of children with lung biopsy or HRCT proven interstitial lung disease and their outcome. We collected detailed history, clinical presentation, laboratory findings, findings on lung biopsy and HRCT findings from case sheets. And we assessed the outcome in terms of morbidity and mortality.*
Results: *Male predominance (60%), persistent cough (100%), Velcro crackles (77.5%) on examination, neutrophils in bronchoalveolar lavage and mean survival period of 2 years and 9 months are the important observations made in our study.*
Conclusions: *Most of the children with ILD presented with chronic cough, dyspnoea and hypoxemia. High resolution Computer tomography appears to be highly sensitive. It appears that after completion of steroid, maintenance with hydroxychloroquine prolongs the life expectancy.*
Key Word: *Interstitial lung disease (ILD), Pediatric, Lung biopsy, HRCT*

Date of Submission: 10-02-2025 Date of Acceptance: 20-02-2025

I. Introduction

Diffuse parenchymal lung disease in children, also called Pediatric Interstitial lung disease, comprise a heterogeneous group of chronic pulmonary disorders of both known and unknown etiologies that share common histological features [1,2,3, 4]. There are very few studies so for been reported on this issue because of its rarity and diagnostic conformation [4]. Hence, a retrospective study was done to analyze the epidemiology, clinical presentation and outcome of children with lung biopsy proven interstitial lung disease.

II. Material And Methods

A retrospective analysis of case records of children with lung biopsy or HRCT proven interstitial lung disease admitted in the pediatric department in a tertiary care teaching institute during 10year period, between Jan 2010 – Dec 2019 was done. Presenting symptoms, duration of illness, risk factors, past history, family history, Clinical findings, treatment and outcome were analyzed. results of serial skiagrams of chest, high resolution computed tomography (HRCT), bronchoalveolar lavage (BAL) and lung biopsy were also evaluated.

III. Result

40 cases were included in the analysis; among them 24 (60) cases were males and 16 (40%) were females with mean duration of symptoms at presentation 12.55 months. 14 (35%) cases had onset of symptoms in infancy, 8 (20%) cases had family history of sibling death due to similar chronic lung disorders. 12 (30%) cases of which 10 were exposed to asbestos and 2 cases to Talc. Persistent cough (100%), Velcro crackles 31(77.5) were the commonest physical findings present in these children.14 (35%) cases had cyanosis, 18 (45%) cases were having clubbing at the time of presentation and 8 (20%) cases presented with pulmonary hypertension. Table No.1 shows demographic and clinical profile of the children with ILD.

Table No. 1: Demographic and Clinical Profile of Children With ILD

Demographic and Clinical profile of cases	No. of cases(n=10)
Sex	
Males	24(60%)
Females	16(40%)
Age	
Infants	14(35%)
>1 year	26(65%)
Signs and symptoms	
Persistent Cough	40(100%)
Velcro crackles	31(77.5%)
Clubbing	18(40%)
Cyanosis	14(30%)
Pulmonary Hypertension	08(20%)

Tuberculosis workup was negative in all 40 cases but 17 (42.5%) cases were treated with anti tuberculous drugs of varying duration before presentation. Serial skiagrams were commonly reported as diffuse infiltrates suggestive of interstitial lung disease in 28cases (70%). HRCT was done in 36 cases of which 31 cases (86.1%) were reported to have features of interstitial lung disease and 5 were reported as bronchiectasis. Ground glass opacification was the commonest abnormality reported in all these31 cases. BAL done in 16 (40%) cases revealed inflammatory cells predominantly neutrophils in addition to macrophages. Open lung biopsy was done in 21(52.5%) cases and histopathological examination of tissue sample revealed interstitial fibrosis with diffuse infiltrates as commonest finding. All patients were started on oral prednisolone 1-2 mg/kg/day with or without hydroxychloroquine. Steroids were continued for at least 3 months and tapered depending upon the response. On follow up, 22 (55 %) cases died due to respiratory failure with mean survival duration of 2 years and 9 months after initial diagnosis. Rest of the children continued to improve with drug therapy.

IV. Discussion

Differential diagnosis of pediatric interstitial lung diseases remains a challenge involving more than a hundred causes [1, 6]These diseases remain difficult to manage, since the small number of cases is a fact that limits the possibilities of clinical studies and research. Though noninvasive techniques are available to diagnose these disorders, lung biopsy remains gold standard[5]. In our study of 40 cases, 14 (35%) cases were below 1 year of age. This presentation in infancy is in accordance with previous studies [1, 4, 7]. Male children were affected more commonly than female in the ratio of 1.5:1 and are similar to other studies [4]. We found 8 cases with history of sibling death due to similar chronic lung disorder, which might suggest genetic predisposition. Exposure to asbestos or excessive use of talc was found in 12 children but their direct causal relationship could not be established even after lung biopsy. Though all children were found to be negative for tuberculosis workup, 42.5% children were treated with anti-tubercular drugs for varying duration before presentation due to endemic nature of tuberculosis in India. On physical examination dyspnoea and Velcro crackles (77.5%) were the most common sign unlike ERS task force study, where only 44% had Velcro crackles[4].Clubbing (40%), cyanosis (30%) and pulmonary hypertension and/or corpulmonale in advanced cases which is consistent with ERS task force study [4]. Most children (80%) had history of two or more episodes of hospitalization for recurrent and chronic respiratory symptoms.

Initial chest radiography was not suggestive of interstitial lung disease but serial skiagrams showed features of ILD. As HRCT was suggestive of ILD in 77.5% cases, this noninvasive technique though not gold standard might be considered a useful tool in diagnosing ILD in children, especially in resource limited settings where lung biopsy is not feasible. Some pediatric literatures confirm that HRCT increases the level of diagnostic confidence for infiltrative lung disease [8, 9]. Ground glass opacification was the commonest abnormality seen in HRCT, a finding which is consistent with most of the previous studies [4,9, 10]

Bronchoalveolar lavage was not done as a routine for assessing the cellularity since it requires child to be sedated and increase the risk of hypoxemia. In our study, BAL was done only in 18 cases which showed predominance of neutrophils, a finding associated with idiopathic pulmonary fibrosis [11, 12]. Although useful tool in diagnosis, BAL cannot replace lung biopsy in the diagnosis of ILD [13]. Lung biopsy was done as open lung biopsy in 21 cases. The predominant histopathological finding in all cases was diffuse fibrosis with inflammatory infiltrates in alveoli and interstitium.

All patients were initially started on oral prednisolone 1-2 mg/kg/day singly or in combination with hydroxychloroquine 10mg/kg/day the most commonly used drug treatment[14].Steroids were continued for at least 3 months and tapered depending upon the response and these children were on follow up. Analysis of readmission records revealed that 22 cases died of respiratory failure with mean survival duration of 2 years and 9 months from the time of diagnosis. The rest of the cases on hydroxychloroquine after tapering the steroids were on regular follow up and on assessment, found to have decrease in dyspnea, better exercise tolerance and

increase in Sao2 at rest. Hydroxychloroquine along with initial steroids appears to be a good regimen[4].However large randomized trials are required to confirm the same.

V. Conclusion

Since HRCT appears to be useful diagnostic tool, almost equal to open lung biopsy it should be done in all children suggestive of ILD where lung biopsy is not feasible. It appears that after completion of steroid, maintenance with hydroxychloroquine prolongs the life of these children with less complication.

References

[1] Fan LL, Deterding RR, Langston C: Pediatric Interstitial Lung Disease Revisited. Pediatrpulmonol 2004, 38:369-378.
[2] Clement A, Eber E: Interstitial Lung Diseases In Infants And Children. Eurrespir J 2008, 31:658-666.
[3] Deutsch GH, Young LR, Deterding RR, Fan LL, Dell SD, Bean JA, Et Al. Diffuse Lung Disease In Young Children: Application Of A Novel Classification Scheme. Am J Respircrit Care Med. 2007;176(11):1120-8.
[4] Clement A, Allen J, Corrin B, Dinwiddie R, Lepointe HD, Eber E, Et Al. Task Force On Chronic Interstitial Lung Disease In Immunocompetent Children. Eurrespir J 2004; 24: 686-697.
[5] Niguidula FN, Balsara RK, Cuasay RS. The Role Of Lung Biopsy In Interstitial Lung Diseases In Children. In: Laraya-Cuasay LR, Hughes WT, Eds. Interstitial Lung Disease In Children, I. Boca Raton, FL: CRC Press, 1988: 121-150.
[6] Bokulic RE, Hilman BC. Interstitial Lung Disease In Children.Pediatrclin North Am 1994; 41: 543-567.
[7] Stillwell P, Norris D, O'Connel E, Rosenow E, Weiland L, Harrison E. Desquamative Interstitial Pneumonitis In Children. Chest 1980; 77: 165-171.
[8] Seely JM, Effmann EL, Muller NL. High-Resolution CT Of Paediatric Lung Disease: Imaging Findings. AJR Am J Roentgenol 1997; 168: 1269–1275.
[9] Copley SJ, Padley SP. High-Resolution CT Of Paediatric Lung Disease.Eurradiol 2001; 11: 2564–2575.
[10] Brody AS. Imaging Considerations: Interstitial Lung Disease In Children.Radiolclin North Am 2005; 43: 391-403.
[11] Osika E, Muller MH, Boccon- Gibod L, Clement A. Idiopathic Pulmonary Fibrosis In Infants. Pediatrpulmonol 1997; 23: 49-54.
[12] Rudd R, Haslam P, Turner-Warwick M. Cryptogenic Fibrosingalveolitis: Relationship Of Pulmonary Physiology And BAL To Responseto Therapy And Prognosis. Am Rev Respir Dis 1981; 124: 1-8.
[13] J. Riedler, J. Grigg, C.F. Robertson Role Of Bronchoalveolar Lavage In Children With Lung Disease. Eurrespir J, 1995, 8, 1725–1730.
[14] Fan L, Langston C. Paediatric Interstitial Lung Disease. Amjrespircrit Care Med 2002; 165: 1466–1467.