

Chronic interstitial lung disease (ChILD) in childhood

M N Lucas¹, H M K N Hathlahawatta², R Seneviratne³, D Samarage⁴

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Introduction

Chronic interstitial lung disease (ChILD) in children is a diverse group of conditions primarily involving the alveoli and perialveolar tissue, leading to a derangement of gas exchange, restrictive lung physiology and diffuse infiltrates on radiographs¹. To date this disorder is found to be rare in children and neither incidence or prevalence is known. This is the first reported case in Sri Lanka.

Case Report

A 1½ year old girl, a product of incest, was admitted on 13th of March, 2007 with a history of cough, cold and shortness of breath of 2 days duration. On examination, she weighed 5kg and had severe failure to thrive, dyspnoea, tachypnoea (respiratory rate 70–80/min), intercostal and subcostal recession, flaring alae nasi, bilateral rhonchi and crepitations. She was treated with intravenous antibiotics for 7 days, oxygen for 10 days and regular nebulisation. She responded slowly and was off oxygen in 10 days. Her lungs were clear but tachypnoea persisted. In 5 days time there was worsening of her symptoms and she developed fever, became ill and oxygen dependant with another respiratory infection. With similar treatment she recovered after about a week but the tachypnoea persisted. There was no contact history of tuberculosis.

Chest X rays done 2 weeks apart showed persistent infiltration of bilateral lung fields (Figures 1 and 2).

¹Registrar, ²Senior Registrar in Paediatrics, Colombo South Teaching Hospital, ³Senior Lecturer & Head, ⁴Senior Lecturer, Department of Paediatrics, University of Sri Jayawardenapura.

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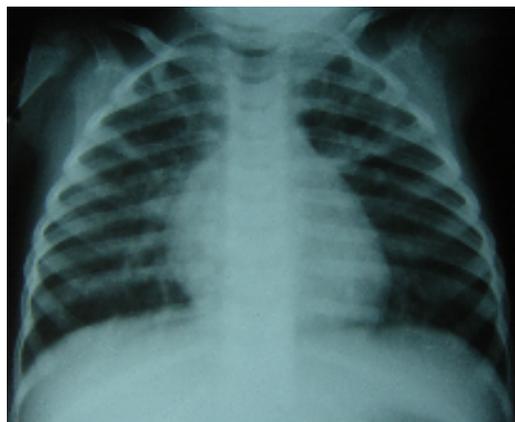


Figure 1 Initial chest x-ray showing bilateral lung infiltration

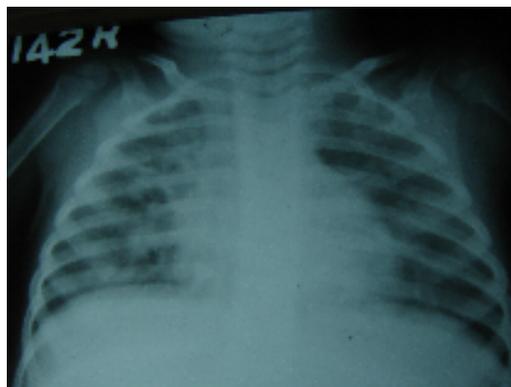


Figure 2 Chest X-ray 2 weeks later showing persistent infiltration

ESR was high (80-85mm 1st hr) at times of exacerbation and normal (6-8mm 1st hr) in between. High Resolution CT (HRCT) scan of chest showed diffuse ground glass appearance with thickened interstitium (Figure 3). Antibody to Human Immunodeficiency Virus was negative. Mantoux test as well as gastric aspirate for acid fast bacilli were negative.

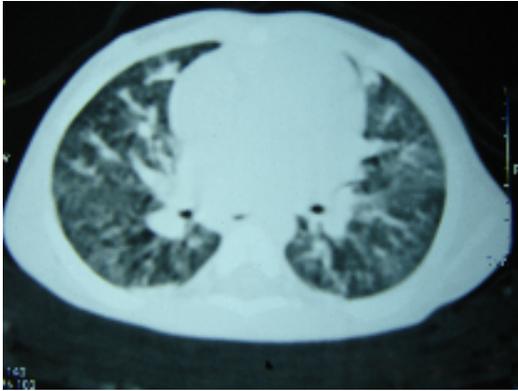


Figure 3 High resolution CT chest showing diffuse ground glass appearance with thickened interstitium

Based on the above chest X ray and HRCT chest findings she was diagnosed to have chronic interstitial lung disease and treatment was initiated with steroids. Treatment was commenced with prednisolone 2mg/kg/day for 2 weeks with minimal response. Therefore pulse therapy with methyl prednisolone was commenced 30mg/kg/day for 3 consecutive days monthly. A good response was seen with lower respiratory rates and better pulse oximetry readings after the first cycle of methylprednisolone (respiratory rate reduced from 70-80/min to 50-60/min and O₂ saturation at rest increased from 90-91% to 94-95%) and she was discharged from hospital. She will be readmitted monthly for pulse therapy for a minimum of 3-6 months.

Discussion

ChILD in immunocompetent children is defined as the presence of respiratory symptoms and/or diffuse infiltrates on chest radiographs, abnormal pulmonary function tests with evidence of restrictive ventilatory defect and/or impaired gas exchange, and persistence of any of these findings for >3 months². It occurs more commonly in young children³. Parental consanguinity is present in about 10% of children⁴. Failure to thrive is documented in almost two thirds of patients diagnosed before the age of 2 years⁴. Chest X-rays and HRCT scans predominantly show interstitial infiltrates⁴. Oral prednisolone (1-2mg/kg/day) or pulsed intravenous methyl prednisolone, singly or in combination with hydroxychloroquine are the most commonly used drug treatments². The natural history of the disease is very variable and some cases burn out spontaneously even without treatment, whilst others progress relentlessly towards a fatal outcome despite all treatment modalities given⁵.

Acknowledgements

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