



British Paediatric Orphan Lung Diseases (BPOLD)

Interstitial Lung Disease - Dr Robert Dinwiddie.

Consultant Respiratory Paediatrician and Honorary Senior Lecturer. Great Ormond Street Hospital for Children and Institute of Child Health. London.

The treatment of interstitial lung disease in children depends on the nature of the underlying pathology. In approximately 50% of cases a specific aetiology can be found such as: chronic viral infection, an auto-immune process, sarcoidosis or alveolar proteinosis. In the remainder, the process is idiopathic and the pathological findings are based on the descriptive morphological features seen in the diagnostic lung biopsy. If a specific cause is found then targeted treatment with antivirals, steroids or other immunosuppressive agents is available. Alveolar proteinosis can be treated by bronchial lavage and GM-CSF. Idiopathic cases are treated primarily with intravenous pulsed methylprednisolone or oral prednisolone backed up hydroxychloroquine. Other immunosuppressive agents such as azathioprine, methotrexate or ciclosporin have been used successfully in individual patients.

The prognosis is very variable and includes no response to any therapy, partial response with chronic long term morbidity, to virtually complete recovery. The overall mortality rate is 15%

There are no controlled therapeutic trials available because of the rarity of these conditions in childhood. Unlike in adult practice, no correlation has as yet been demonstrated between the initial pattern of chest x-ray change or the degree of pathological change on the lung biopsy and the clinical outcome.

The recurrence rate within families is 1 in 8.

From Paediatric Respiratory Reviews (2004) 5, 108-115. Copyright Elsevier Ltd.

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See also:

Fibrosing Alveolitis and Desquamative Interstitial Pneumonitis	N. Sharief, O.F. Crawford, R. Dinwiddie	Pediatr Pulmonol. 1994; 17:359-365
Interstitial lung disease in children: a multicentre survey on diagnostic approach	A. Barbato, C. Panizzolo, A. Cracco, J. de Blic, R. Dinwiddie, M. Zach	Eur Respir J 2000; 16:509-513
Idiopathic Interstitial Pneumonitis in Children: A National Survey in the United	R. Dinwiddie, N. Sharief, O. Crawford	Pediatr Pulmonol. 2002; 34:23-29



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