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Understanding Pulmonary Fibrosis in Pediatric Interstitial Lung Disease:A Comprehensive Analysis

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Abstract

Introduction We aimed to investigate the prevalence of pulmonary fibrosis in chILD and its impact on disease prognosis using data from the chILD-Turkiye (chILD-TR) registry.

Method All center registered in the chILD-TR in 2024 were included in the study. Thoracic CT scans were reviewed by an experienced radiologist for pulmonary fibrosis findings. The Warrick ILD score were assessed. The clinical and demographic data of the patients were analyzed.

Results A total of 404 patients (47.5% girls) were included, median age was 137 (24-376) months. The median weight z-score was -1.02 (-8.8-6.88), height z-score was -0.59 (-7-6.45). Median FEV1% was 68 (16-127) and FVC% 64 (17-136), and DLCO was 66% (21-132). The pulmonary fibrosis findings were as follows: reticular abnormalities in 34.9% of patients, cystic abnormalities in 21%, traction bronchiectasis in 19.8%, architectural distortion in 14.4%, and honeycombing in 2.5%. Additional findings included ground-glass opacity in 83.6% of patients, mosaic perfusion in 41.9%, nodules/nodular opacities in 27.5% and pleural irregularity 19%. Two groups were compared based on the presence (n=183) or absence (n=221) of pulmonary fibrosis findings, the group with fibrotic findings had a higher age and lower weight z-score (p<0.05). While no differences were found in PFT parameters between the groups (p>0.05), DLCO% was lower in the fibrotic group (p=0.015). The median Warrick severity score was 3 (0-15), extended score was 3 (0-15) and the Warrick total score was 6 (0-30).

Conclusion Pulmonary fibrosis is a common complication in chILD and becomes more prominent with age. The association of fibrosis with older age and lower DLCO % highlights the need for early detection.

Footnotes

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The Importance of Bronchoalveolar Lavage Findings in Diagnosis and Their Relationship with Radiological Findings in Pediatric Interstitial Lung Diseases: Turkey chILD National Registry Cohort Study

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Abstract

Introduction: Bronchoscopy and cell count analysis in interstitial lung diseases (ILD) provide valuable information at the alveolar level. Computed tomography (CT) is considered the gold standard for characterizing ILD. The aim of this study is to evaluate the diagnostic significance of bronchoalveolar lavage (BAL) findings and their relationship with radiological findings in ILD patients from the National Registry System. **Materials and Methods:** Data from patients registered in the National Registry System between 2022 and 2023 were retrospectively reviewed. Disease subgroups, serological and radiological findings, and BAL results were analyzed. **Results:** A total of 599 patients were registered, of which 255 had undergone bronchoscopy and chest CT. When disease diagnoses were categorized, the most frequent diagnosis was B2 (immune function presumed normal in the host) in 70 (27.4%) patients. The most common radiological finding was ground-glass opacity, seen in 151 (61.4%) cases. Specific diagnoses included pulmonary alveolar proteinosis

(PAS+ in 6 [2.4%] patients. When the relationship between BAL findings and radiological findings was assessed, a significant association was found between increased levels of hemosiderin-laden macrophages and interstitial thickening. A significant correlation was also observed between interstitial thickening and between focal consolidations.

Conclusion: The cytological evaluation of BAL, when used in conjunction with chest CT, can provide valuable diagnostic insights in ILD and may reduce the need for invasive procedures such as surgical lung biopsy.

Footnotes

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Quality of Life in Turkish Children with Interstitial Lung Disease: Insights from the National ChILD Registry

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Abstract

Introduction: This study, the first of its kind in this population, evaluates health-related quality of life (HRQoL) in children with childhood interstitial lung disease (ChILD) and their families using data from the Turkish National ChILD Registry.

Methods: As of January 2024, data from 17 centers were analyzed. Patients aged 1 month to 18 years and their families completed the Pediatric Quality of Life Inventory (PedsQL™ 4.0 Generic Core Scales) and PedsQL™ 2.0 Family Impact Module through face-to-face interviews. Higher scores indicate better HRQoL. The patients' PFT and Fan scores were recorded.

Results: The study included 175 patients (90 girls), with a median age of 93 (13–214) months. FEV₁% positively correlated with the PedsQL™ Physical Health Summary Score, Parent-Reported Physical Health Summary Score, Psychosocial Health Summary Score, total score ($p=0.007$, $r=0.608$; $p=0.025$, $r=0.527$; $p=0.032$, $r=0.522$; $p=0.009$, $r=0.612$). The Fan score negatively correlated with

PedsQL™ scores ($p < 0.001$). No significant correlation was found between FEV₁% and the Family Impact Module total score, though a weak negative correlation with the Fan score was noted ($p < 0.001$ – 0.01).

Conclusion: Lung function strongly correlates with HRQoL, emphasizing the impact of disease severity on physical and psychosocial well-being. These findings highlight the need for multidisciplinary care and psychosocial support to enhance the quality of life in children with ChILD and their families.

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