European Respiratory Society
Annual Congress 2013

Abstract Number: 4815

Publication Number: P1246

Abstract Group: 7.7. Paediatric Bronchology

Keyword 1: Infections **Keyword 2:** Bronchoalveolar lavage **Keyword 3:** Bronchoscopy

Title: Interstitial lung diseases in children in a tertiary hospital in North Italy

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Body: Interstitial lung diseases (ILDs) in children are an heterogeneous group of lung disease with very different pathophysiology. In most European countries rare orphan diseases such as pediatric interstitial lung diseases (ILDs) are still underdiagnosed. Objective: To investigate/ describe pediatric ILDs in a tertiary university hospital in North Italy. Methods: we included 18 children admitted to the Pediatric Department of the Padova University Hospital from 2003 until 2012 with a diagnosis of ILD. Children were identified retrospectively through clinical charts and clinical, radiological, functional, pathological, biological and genetic data were collected. Results: Data were available for x children presenting with ILD. Median age at diagnosis was 5.5 years [1-16 years], and the sex ratio was 0.5 male/female. Radiologic, laboratory histology and genetics led to the following diagnosis: Bronchiolitis obliterans organizing pneumonia (BOOP), surfactant protein B mutation and viral induced interstitial lung disease were the most common diagnoses. At high resolution CT-scan, 14/18 children showed in 16 % a Honey comb pattern, in 16 %, a reticular picture, in 16% a nodular picture, and in 52% ground glass opacity. Chronic cough and wheezing episode were the most common symptoms reported. Conclusion: In Italy, pediatric ILD is still under estimated. A national database, such as the German, UK or France registry, and national guidelines for diagnosis and possible therapies are necessary to allow physicians and patients to have a more adequate diagnosis and follow-up.