

# 8<sup>th</sup> Molecular Immunology & Immunogenetics Congress

March 20-21, 2017 Rome, Italy

## Distinct effect of interleukin 4 and B-cell activating factor in connective tissue diseases with lung involvement and idiopathic pulmonary fibrosis

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**Statement of the Problem:** In patients with connective tissue diseases (CTDs) lung involvement may play an important prognostic role. Possible radiologic pattern of lung involvement includes usual interstitial pneumonia, typical also for idiopathic pulmonary fibrosis (IPF). We hypothesize that despite the same radiologic pattern the pathogenetic pathways in IPF and CTD may differ.

**Methodology & Theoretical Orientation:** Sixteen naive CTD patients with lung involvement according to high resolution tomography of the chest and 17 IPF patients were included and underwent serum Ig concentration assessment, flow cytometry of peripheral blood and bronchoscopy with bronchoalveolar lavage (BAL). BAL fluid (BALF) differential cell counts and measurement of IL-4R and BAFF in BALF supernatant were performed.

**Findings:** CTD patients exhibited higher BALF lymphocyte counts ( $p < 0.05$ ) and lower IL-4R concentrations ( $p < 0.05$ ). BAFF concentrations did not significantly differ between groups. Peripheral blood lymphocyte and CD19 counts were higher in IPF patients ( $p < 0.05$ ). Autoantibodies were positive in all CTD and in 8 IPF patients. BAFF concentrations positively correlated with IgM serum concentrations (both in CTD and IPF patients,  $p < 0.05$ ). Positive correlations among IL-4R concentrations, serum IgG, IgA and IgM concentrations and CD19 peripheral blood counts ( $p < 0.05$ ) were observed in both groups. Correlations between BAFF and IL-4R BALF concentrations were positive in CTD ( $p < 0.01$ ) and negative in IPF patients ( $p < 0.05$ ).

**Conclusion & Significance:** Our data suggest that the immunologic and molecular markers in peripheral blood and BALF may differ in IPF and CTD patients. We hypothesize that anti-apoptotic effect of IL-4 on B cells may be clinically relevant in IPF patients, while different regulatory pathway may play a role in CTD.

### Biography

Martina Sterclova has been working since 2004 as a Medical Doctor in the Department of Respiratory Diseases, Thomayer Hospital, Prague. In 2009, she finished her PhD studies in Immunology and Molecular Biology. Her area of interest concerns interstitial lung diseases, namely role of cytokines and chemokines in disease pathogenesis. In 2013, she gained ERS Fellowship and studied bronchoalveolar lavage cell cultures at Comprehensive Pneumology Center, Munich, Germany. She regularly publishes and actively participates in various international conferences on interstitial lung diseases.

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