388. Diffuse parenchymal lung disease II

P3630
Hypersensitivity pneumonitis by feather duvet: A series of Vall d’Hebron Hospital

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Introduction: Exposure to avian proteins is a frequent cause of Hypersensitivity Pneumonitis (HP). We describe a series of NH secondary to exposure to feather duvet.

Material and methods: In the outpatient clinic of interstitial lung disease during the years 2004-2011, 28 patients were diagnosed with HP with definite causality from exposure to feathers contained in the feather duvet. Diagnosis criteria: diagnosis of NH, contact with feather duvet coincident with the onset of symptoms; IgG+ and/or positive bronchial challenge test (BCT). In neither case had history of exposure to any agent known as a producer of NH.

Results: 15 male; mean age 59 years. Presentation was acute in 5, subacute in 4 and chronic in 19 cases. Auscultation was normal in 10 patients, revealed crackles in 15 and wheezing in 1. FVC mean was 67.57%, DLCO 52.60%. IgG+ in front of avian antigens in 11/24, and in front of fungi in 16/24. A culture of feather duvet was positive for fungi in 6 cases. The BCT was positive in 8/11 in front of avian antigens and 4/7 against fungi. Pathological study was performed in 16/28 patients, showing a characteristic pattern of subacute NH in 6/16, consistent with NH in 8/16 and UIP pattern in 2/16. Surgical lung biopsy (BQ) was performed during the study in 7 cases and revised from the samples of another center in 7.

Conclusion: Our study confirms that exposure to a minimum but persistent agent may be sufficient for disease development. Diagnosis at an advanced stage of chronic NH is common. Project funded by FIS PI1001577 (ISCIII) and SEPAR 2010.

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P3631
Macrolides inhibit cytokine production by alveolar macrophages in bronchiolitis obliterans organizing pneumonia

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Background and objective: Bronchiolitis obliterans organizing pneumonia (BOOP) is a distinct clinicopathological entity histologically characterized by intra-alveolar granulation tissue and absence of extensive fibrotic lesions. Effective macrolide treatment of BOOP has been reported anecdotally. This study aimed to investigate whether alveolar macrophages (AMs) produce aberrant proinflammatory cytokines in BOOP and whether this can be inhibited by clarithromycin (CAM) or azithromycin (AZM).

Methods: AMs collected by bronchoalveolar lavage (BAL) from 6 BOOP patients and 8 non-ILD controls were cultured for 24 hours in the presence or absence of CAM, AZM, lipopolysaccharide (LPS), or dexamethasone (DEX). Tumor necrosis factor alpha (TNF-α), soluble TNF receptor 1 (sTNFR1), sTNFR2, interleukin 1β (IL-1β), IL-6, IL-8, IL-10, interferon gamma inducible protein 10 (IP-10) and CC chemokine ligand 18 (CCL18) were measured in the culture supernatant by ELISA.

Results: The spontaneous and LPS-stimulated production of all investigated cytokines by AMs was significantly increased in BOOP compared to controls. CAM and AZM induced a dose-dependent suppression of spontaneous TNF-α, sTNFR2, IL-6, IL-8 and CCL18 production (p < 0.05). CAM also inhibited the IL-1β production. CAM and AZM significantly and dose-dependently attenuated the LPS-stimulated production of sTNFR1, sTNFR2, IL-8 and CCL18 (p < 0.05). CAM also inhibited the LPS-stimulated TNF-α, IL-1β, IL-6 and IL-10 production.

Conclusions: AMs from BOOP patients produce abundant proinflammatory cytokines which may be pivotal in the disease pathogenesis. Macrolides inhibit this cytokine production, CAM more efficiently than AZM.

P3632
Comparison of clinical features between patients of lung cancer with combined pulmonary fibrosis and emphysema syndrome and those with interstitial pneumonitis

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Purpose: Patients with combined pulmonary fibrosis and emphysema syndrome (CPFE) and those with interstitial pneumonitis (IP) have a high risk of developing lung cancer. The present study compared the clinical features of patients with lung cancer and CPFE with those of patients with lung cancer and IP.

Methods: 48 patients with lung cancer and CPFE (CPFE group) and 30 patients with lung cancer and IP (IP group) had surgery and were included this study.

Results: All patients in the CPFE group and 24 of 30 patients in the IP group were current or ex-smokers (p=0.01) (Table 1). The CPFE group tended to have more patients with IPF than the IP group (p=0.09). FEV1.0% predicted was significantly lower in the CPFE group than in the IP group (Table 2). Acute exacerbations of
interstitial lung disease after surgery developed in three patients (6.3%), one of whom died, in the CPFE group and two patients (6.7%) in the IP group (p=0.94).

### Table 2. Laboratory findings and pulmonary function tests

<table>
<thead>
<tr>
<th>Patients with lung cancer and CPFE</th>
<th>Patients with lung cancer and IP</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>(n=40)</td>
<td>(n=30)</td>
<td></td>
</tr>
<tr>
<td>PaO2 (torr)</td>
<td>85.0±11.3</td>
<td>85.0±10.8</td>
</tr>
<tr>
<td>A-aDO2</td>
<td>13.1±10.9</td>
<td>15.2±11.5</td>
</tr>
<tr>
<td>Laboratory</td>
<td></td>
<td></td>
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<tr>
<td>K-L (6-Unit)</td>
<td>534.0±240.9</td>
<td>764.0±458.2</td>
</tr>
<tr>
<td>SP-D (pg/l)</td>
<td>114.6±73.2</td>
<td>128.5±74.7</td>
</tr>
<tr>
<td>SP-A (mg/l)</td>
<td>0.9±0.6</td>
<td>0.9±0.7</td>
</tr>
<tr>
<td>LLl (l)</td>
<td>1.8±0.4</td>
<td>2.0±0.5</td>
</tr>
<tr>
<td>LHFi (pg/ml)</td>
<td>5.1±1.5</td>
<td>6.2±1.4</td>
</tr>
<tr>
<td>Pulmonary function tests</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VC % pred</td>
<td>102.0±18.5</td>
<td>99.5±16.2</td>
</tr>
<tr>
<td>FEV1 % pred</td>
<td>93.5±16.4</td>
<td>99.2±11.2</td>
</tr>
<tr>
<td>TLC % pred</td>
<td>98.0±12.4</td>
<td>98.0±8.8</td>
</tr>
<tr>
<td>RV % pred</td>
<td>94.5±26.2</td>
<td>90.0±21.1</td>
</tr>
<tr>
<td>DCO % pred</td>
<td>63.0±12.3</td>
<td>86.0±11.0</td>
</tr>
</tbody>
</table>

Data are presented as mean ±2SD

Conclusions: Although IPF was more frequent in the CPFE group than in the IP group, the occurrence of acute exacerbations after surgery was not different between the two groups.

### P3633

A case of lymphoid interstitial pneumonia associated with common variable immunodeficiency

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**Affiliations:** 1 2 3 4 5 6

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**Introduction:** Lymphoid interstitial pneumonia (LIP) represents a rare disease that typically occurs in association with autoimmune diseases and dysproteinemia. We report a case of LIP in a patient with common variable immunodeficiency (CVID).

**Case report:**

A 51 year old woman presented with a 12 months history of recurrent pyrexia and was subsequently diagnosed with CVID. CT scans showed bilateral pulmonary consolidations, while bronchoscopy revealed acute inflammation of the bronchial mucosa and purulent secretion. The patient received antibiotic treatment and immunoglobulin replacement. She remained asymptomatic until January 2012, when she was readmitted with increasing dyspnea and fatigue for 2 weeks prior to admission. CT scans showed progressive opacities mainly in the lower zones. Despite immediate treatment with broad-spectrum antibiotics, the patient developed acute respiratory failure requiring invasive mandatory ventilation and, ultimately, extracorporeal membrane oxygenation. Open lung biopsy was performed 5 days after admission. Histologic workup showed diffuse interstitial infiltration of T lymphocytes, plasma cells and histiocytes, consistent with a diagnosis of LIP.

**Discussion:** The finding of bilateral ground-glass infiltrates in a patient with CVID should raise suspicion of LIP. Open lung biopsy is required to establish the diagnosis. Treatment is primarily based on corticosteroids, which resulted in excellent clinical response in this patient.

### P3634

Disease progression according to IPF phenotype

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**Introduction:** Our understanding of disease progression in IPF is based on cohorts of patients with 'definite' IPF. Such studies have reported a prognostic index for IPF (De Bois 2011 ARCCM 184; 459). In practice a diagnosis of definite IPF is not always attained; in some cases where the HRCT pattern is not incontrovertibly UIP, corroboration by invasive investigations is not performed. We define this condition as "probable IPF.

**Aims:** To determine 1) disease progression and 2) the prognostic value of an IPF index in definite or probable IPF.

**Methods:** Consecutively presenting patients with IPF were prospectively recruited to a database. All IPF diagnoses required an HRCT appearance of UIP with >70% probability. Patients with HRCT scans with >95% probability of UIP or a UIP biopsy were defined as definite IPF. BAL was not included in the diagnostic criteria.

**Results:** Of 193 patients, 89 had definite IPF. Median survival for definite v
Thematic Poster Session

P3638
Natural history of idiopathic pulmonary fibrosis: Are slow progressors really steady conditions?
Elisabetta Balestico, Emanuela Rossi, Ariel Fioriliano, Claudia Rinaldo, Francesca Lunardi, Monica Loy, Federico Rea, Manuel Cosio, Marina Saetta, Fiorella Calabrese, Department of Cardiologie, Thoracic and Vascular Sciences, University of Padova Medical School, Padova, Italy

Idiopathic pulmonary fibrosis (IPF) is a devastating lung disease with heterogeneous clinical course. Some patients experience an accelerated disease progression (rapid progressors) while other remain relatively stable over time (slow progressors).

The aim was to investigate the different course of the disease in relation to survival. The study population included 55 IPF patients (age at diagnosis 53±11) categorized in rapid progressors and slow progressors by two distinct criteria: pre-diagnosis criteria (decrease in lung function tests) and post-diagnosis criteria (decline in FVC±10% over 12 months). Stratified survival analysis was performed using the Kaplan-Meier method.

Results: Using post-diagnosis criteria, 18% were considered in the group of slow progressors while 66% were fast progressors. Median survival was 4.7 years for slow progressors and 2.5 years for fast progressors. The survival curves showed significant differences between the two groups (log-rank test p<0.001). Of interest, rapid progressors according to post-diagnosis criteria had a prognostic significance; indeed, rapid progressors had decreased survival as compared to slow ones (28.4±15.4 v.s. 39.1±15.6). From these data, it can be concluded that IPF patients have a variable and unpredictable clinical course rather than a steady condition.

In conclusion, we suggest that IPF patients to be categorized in the beginning of the disease. The identification of these patients is crucial for the early intervention and management of the disease.

P3639
The clinical relevance of autoimmunity in idiopathic pulmonary fibrosis
Sara Tomassetti, Christian Guirol, Claudia Ravalga, Sara Picciocchi, Gian Luca Casini, Micaela Romagnoli, Carlo Guirol, Venerino Polletti. Pulmonary, GB Morgagni Hospital, Forli, Italy

The aim of this study is to evaluate the clinical significance of autoimmunity in IPF:

1. This is a retrospective controlled study comparing clinical characteristics and outcome of IPF patients with (n=73) and without (n=87) positive autoimmunity. Seventy-three cases with positive autoimmunity included: 46 (63%) ANA (anti-nuclear antibody), 20 (27%) RF (rheumatoid factor), 3 (4%) ENA (antibodies to extractable nuclear antigens). 2 (3%) ANCA (anti-neutrophil cytoplasmic antibody).

2. There were no differences between patients with positive and negative autoimmunity in terms of age, gender, smoking history, prevalence of familial IPF, high resolution computed tomography features.

Abstract P3640 – Table 1. Correlations between radiographic stages, HRCT scores and subscores and functional parameters.

<table>
<thead>
<tr>
<th>Parameters</th>
<th>FEV1</th>
<th>FVC</th>
<th>DLCO</th>
<th>MMEF25-75</th>
<th>TLC</th>
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<tbody>
<tr>
<td>TLC</td>
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<td>FEV1</td>
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<td>FVC</td>
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<td>DLCO</td>
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<td>MMEF25-75</td>
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*P<0.05, **P<0.001

P3640
Sarcoidosis and autoimmune thyroid disease
Claudia Ravalga, Carlo Guirol, Gian Luca Casoni, Micaela Romagnoli, Sara Tomassetti, Christian Guirol, Venerino Polletti. Pulmonology, Department of Thoracic Diseases, Morgagni Hospital, Forli, Italy

Background: Association between Sarcoidosis and other autoimmune diseases has been previously described and a common pathogenesis has been hypothesized.

Objectives: A descriptive retrospective study has been conducted to evaluate the incidence of thyroid diseases in patients with pulmonary sarcoidosis.

Methods: We conducted a retrospective chart review of all patients diagnosed with sarcoidosis between 2004 and 2011 at the Morgagni Hospital, Forli; those who were also diagnosed as having an autoimmune thyroid disease were selected.

Results: 39 out of 246 (15.9%) patients with sarcoidosis were identified as having autoimmune thyroid disease. 9 male and 30 female. Sarcoidosis presented as Löfgren’s syndrome in 6 patients patients (15.4%). 3 patients developed Graves’ disease (7.7%), 12 patients developed Hashimoto’s thyroiditis with hypothyroidism (30.8%) and 24 patients had thyroid nodules with normal thyroid function (61.5%).

Conclusions: Our study suggests that sarcoidosis may be associated with autoimmune thyroid diseases at some point of its evolution, either as hyperthyroidism or hypothyroidism. We are now following up sarcoidosis patients with no thyroid disease to see if any thyroid disease can develop after the diagnosis or during the period of activity of sarcoidosis.

P3641
Assessment of disease severity using imaging scores in pulmonary sarcoidosis
Victoria Botsman, Diána Calárga, Oxana Munetru. Internal Medicine, State Medical and Pharmaceutical University "Nicolaie Testemitanu", Chisinau, Republic of Moldova

The value of imagistic methods in diagnosing and assessing the inflammatory activity in sarcoidosis is well known. The aim was to assess the association between imagistic findings (radiographic score, and HRCT score) and disease severity defined by functional parameters.

We evaluated the clinical records of 70 patients with biopsy proven or highly suggestive of sarcoidosis, cases registered in a 3rd level medical institution, during year 2011, who underwent chest X-ray, HRCT, and pulmonary function tests. The HRCT images were scored by two readers. Spearman’s rank correlation coefficients were calculated to estimate the association between imagistic scores and respiratory function disturbances.

We found a relationship between both radiological staging and HRCT abnormalities with functional parameters. All HRCT subscores, except lymph node enlargement, correlated with FEV1, FVC, MMEF25-75, and DLCO (p<0.001).

Compared with radiological stages, HRCT findings appeared to be more sensitive in tracing abnormal gas exchange, with no significant difference in other functional parameters.
P3642 Myocardial mechanics for the early detection of cardiac sarcoidosis
John Felekos1, Elias Gialafos1, Vasileios Konstantis1, Elka Perros1, Anastasios Kalhanos2, Durais Amangopulos2, George Kaltsakas2, Aggeliki Rapti3, George Tzilepis4, Konstantina Aggelii, 1Cardiology Unit, University of Athens, Greece; 2rd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 3Cardiothoracic surgery, General Hospital of Nikaia, Athens, Greece; 4rd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 51st Pulmonary Clinic, University of Athens, Greece; 6Department of Therapeutics, University of Athens, Greece

Background: Speckle tracking has emerged as valuable tool for comprehensive assessment of regional myocardial function, providing angle-independent measurements of ventricular strain. Aim of this study was to evaluate local ventricular (LV) function in patients with newly diagnosed sarcoidosis, utilizing the novel method of 2D speckle tracking.

Methods: Forty one patients with sarcoidosis and unremarkable medical history of cardiovascular disease, as well as 20 healthy age- and gender-matched controls underwent echocardiographic study. Apical 4-, 2-, 3- chamber as well as short axis acquisitions were made. In addition to conventional 2D, Doppler and TDI measurements, speckle tracking echocardiography was applied and LV global longitudinal strain was derived from the obtained images. Moreover, LV base and apex rotation angles were assessed from which LV twist was derived.

Results: The mean age of patients (17 men) was 41.6±5.0 years old. Compared with controls, patients had similar conventional 2D and Doppler measurements. TDI revealed increased E/E' in the patient group vs control group (8.7±2.1 vs 4.6±2.3, p<0.05). Strain analysis demonstrated reduced global longitudinal strain values in the patient vs control group (18.6±6.1% vs 21.8±8.2%, p<0.05). Furthermore, LV twist was increased in the patient group as compared to the healthy controls (15.2±2.6° vs 10.2±2.1°, p=0.05).

Conclusions: Speckle tracking echocardiography revealed impaired strain and rotational indices, implying elevated filling pressures of the left ventricle. This may represent a sign of myocardial involvement in patients with sarcoidosis. Therefore deformation imaging could be valuable adjunct for screening.

P3643 Overexpression of angiotensin II type 1 receptor (AGTR1) and lymphatic vasculature rarefaction is present in scleroderma with pulmonary compromised
Aline Domingos Pinto Ruppert, Vera Capelozzi, Edwin Roger Pinto, Pathology, Faculdade de Medicina da Universidade de Sã o Paulo, Brazil

Background: The aim of this study was to evaluate the activity of angiotensin II type 1 and 2 receptors and lymphatic vessels in lungs of patients with systemic sclerosis comparing to normal lung tissue (NLT).

Methods: Lungs specimens were obtained from 23 female patients with scleroderma. NLT were obtained from 10 individuals who died suddenly of non-pulmonary causes. Immunohistochemistry and histomorphometry were used to evaluate the amount of AGTR-1 and AGTR-2 pulmonary expression. We also studied the lymphatic (D2-40) expression and the percent of area occupied by these lymphatics in lung specimens. Pulmonary fibrosis was obtained from high-resolution computed tomography (HRCT) score.

Results: We observed higher amount of AGTR-1 and AGTR-2 expression in the patients group than the NLT group (p=0.001). Patients with severe fibrosis showed a increased expression of AGTR-1 and increased area occupied by lymphatic than minimal fibrosis (p<0.05) patients.

Conclusions: Our data showed that the AGTR-1 and AGTR-2 and lymphatic system are very affected in pulmonary SSc. AGTR-1 showed higher expression in more pulmonary fibrosis HRCT score. Rarefaction of lymphatic vasculature was correlated with degree of pulmonary fibrosis and pulmonary function test. The role of AGTR-1 and AGTR-2 are probably essential in evaluation and progression of Systemic Sclerosis.

Financial support: FAPESP

P3644 The relationship between the shuttle walk test, lung function and BMI in patients with sarcoidosis
Robert J Broek, Colin Leonard, Jayne Holme, Annette Duck, North West Lung Centre, University Hospital of South Manchester NHS Foundation Trust, Manchester, United Kingdom

Introduction: Monitoring patients with sarcoidosis is important and challenging. The incremental shuttle walk test is used in outpatient settings and we assessed its correlation with lung function, BMI and age.

Methods: A retrospective study of data from 63 patients with sarcoidosis who had had a shuttle walk with their lung function tests. Spearman’s correlation coefficient r_s was used to assess the relationship between the shuttle walk (distance walked, oxygen saturations) and Borg score (before and after exercise), with lung function (FEV1, FVC, DLCO% predicted), age and BMI. A stepwise multiple linear regression model of distance walked was developed.

Results: Correlation is seen between the shuttle walk and lung function (table 1)The distance walked also correlated with age (r_s=-0.372, p=0.003) and BMI (r_s=-0.303, p=0.016). There was correlation between the Borg score post exercise and the BMI (r_s=-0.253, p=0.046). The multiple regression model of distance walked identified BMI (t=3.195, p=0.002), age (t=2.963, p=0.002) and FEV1 (t=3.061, p=0.003) as significant independent variables.

Conclusion: correlation between shuttle walk test and lung function

P3645 Intestinal lung disease associated with autoimmune thyroiditis (ILD-AT)
Yoko Takahashi, Hiroshi Kudo, Toshikazu Kimura, Kenku Katsaka, Respiratory and Allergy, Tseu General Hospital, Seto, Aichi, Japan

Objective: It has been reported clinical correlation of pulmonary hypertension(PH) in collagen vascular disease(CVD), however the role of PH in intestinal pulmonary related CVD(CVID-IP) has been scarcely evaluated. We sought to determine the prediction factor of mean pulmonary artery pressure(MPAP) in patients with CVID-IP.

Method: Patients with CVID-IP underwent right heart cathereterization(RHC) within 3 months of initial evaluation at our institution. Patients with left ventricular dysfunction, incomplete follow-up, and patients with respiratory failure were excluded. Results: We studied 44 patients, 19 male, mean age 59.8±11.0 years). They were 13 with RA, 13 with SSc, 9 with PM/DM, and 9 with others. PaO2 at rest was 83.1±6.9mmHg, MPAP was 17.2±4.5mmHg (<20mmHg, 13.2±9.5%), cardiac index(DCI) was 3.6±0.8L/min/m2, pulmonary vascular resistance index(PARI) was 223±102 Wood units/m2, ventilative capacity(VC) was 78.4±21.0%, percentage of carbon monoxide diffusion capacity (9DLco) was 50.9±17.6%, and 6-minute walking distance(MWD) was 502±142 m, and minimum SpO2 at 6-minute walking test (mSpO2) was 85.6±5.9%, respectively. The median observation period was 31.8 months with 11 patients died. In the univariable model, MPAP was significantly correlated with 9DLco (r=0.377, P=0.013), and mSpO2 (r=0.552, P<0.001). In the multivariable model, MPAP was significantly correlated with 6MWD (r=0.360, P=0.040) and mSpO2 (r=0.595, P<0.001).

Conclusion: 6MWD and mSpO2 were independent predictors of MPAP in CVID-IP.
P3647
Sarcoidosis – Diagnostics, prognosis and therapy in everyday pneumological practice. A retrospective analysis
Martina Douzhkova, Ilona Binkova, Jana Skrickova. Department of Pulmonary Medicine, University Hospital, Brno, Czech Republic

Background: Currently, prognostic factors, or therapy in sarcoidosis are frequently discussed, however, up-to-date data concerning the incidence and the management of sarcoidosis in everyday pneumological practice are still missing. Therefore, we decided to analyze the characteristic of comprehensive cohort of sarcoidosis patients.

Methods: We retrospectively analyzed all 169 patients, whom the diagnosis of sarcoidosis was set at our pulmonary department in years 2005-2010.

Results: Median age of sarcoidosis patients was 48 years (20-79). Females:males ratio was 1.5:1. Non smoker:smoker ratio was 2.2:1. Familial occurrence was observed in 4 patients. At diagnosis, stage 0 was present in 3.5% patients, stage I in 34% patients, stage II in 49.5% patients, stage III in 11% patients, and stage IV in 2% patients. In 76 patients there was extrapulmonary sarcoidosis. The coincidence of sarcoidosis with other autoimmune diseases was observed in 10 patients; 6 patients developed thrombembolic disease. Spontaneous resolution was seen in 36% patients; 57% of stage I, 29% of stage II, and 11% of stage III. 64% received corticosteroids. 16% of patients developed chronic sarcoidosis. In sarcoidosis patients with spontaneous resolution, no relapse of disease was observed. On the other hand, 10% of treated patients relapsed. Median time to sarcoidosis relapse was 6 months. There was no difference in age between good and poor risk groups of patients. Observed lethality was 1.2% (2 patients).

Conclusion: Male gender, X-ray stage I or II, lymphocytic alveolitis in bronchoalveolar fluid, spontaneous regression, and active disease duration up to 2 years appear to be good prognostic factors.

P3648
BALF cell expression phenotypes and cytokine levels in BALF supernatant in fibrosing interstitial lung diseases
Martina Vasakova1, Ekostra Stancka2, Petra Mandakova1, Martina Stercova1, Jaromir Kaikal1, Radoslav Matej2. 1 Department of Respiratory Medicine, 1st Medical School, Charles University and Thomayer Hospital, Prague, Czech Republic; 2 Department of Pathology and Molecular Medicine, 2nd Medical Faculty, Charles University and University Hospital in Motol, Prague, Czech Republic

Aims: Fibrosing interstitial lung diseases (fILDs), mainly idiopathic pulmonary fibrosis (IPF) and fibrotic nonspecific interstitial pneumonia (NSIP), are processes with unknown etiology and grim prognosis. We hypothesize that expression profile of bronchoalveolar lavage fluid (BALF) cells and BALF cytokine concentrations in fILDs depend on Th2 cytokine milieu and enhance fibroproliferative healing.

Methods: 10 patients with fILDs and 9 controls (C) were included into the study. BALF samples were investigated using FACS to evaluate spontaneous and stimulated production of TNF alpha, percentage of lymphocytes expressing IL-2, IFN-gamma and TNF-alpha and percentage of epithelial cells expressing proteinase-activated receptors (PAR 1,2,3). IFN-gamma, IL-1 beta, IL-2, IL-4, IL-5, IL-6, IL-8, IL-10, IL-12p70 and TNF beta values were evaluated by means of bead-based analysis. Concentrations of CD124, PAR-2, TGF-beta 1 and TNF-alpha in BALF were detected by ELISA as well. Statistical methodology: The differences in basic and derived characteristics were tested via standard two-sample two-sided t-test with correction by False Discovery Rate (FDR) methodology.

Results: We observed significantly higher concentrations of IL-4R and TGF-beta 1 in BALF in fILDs versus C. The expression of PAR-2 on EP was higher in fILDs compared to C as well.

Conclusion: Our results support a hypothesis about the role of Th2 type immune response in pathogenesis of fibrotic ILDs. The higher concentrations of IL-4R might be a sign IL-4 influence, and the increased expression of TGF-beta and PAR-2 gives evidence for ongoing epithelial- mesenchymal transition and fibrogenesis in fILDs.

P3649
Pulmonary eosinophilic syndromes characterization. The role of peripheral eosinophilia as a marker of clinical evolution
Heider Novas e Bastos1,2, Patricia Mota1, Natália Melo1, Marília Beltrão1, Okaina Sokhatska1,2, Eliska Stranska2, Petra Mandakova1, Martina Stercova1, Jaromir Kaikal1, Radoslav Matej2. 1 Department of Respiratory Medicine, 2nd Medical School, Universidade do Minho, Braga, Portugal; 2Escola de Ciências da Saúde, Universidade do Minho, Braga, Portugal

Introduction: Pulmonary eosinophilic syndromes (ES) are a heterogeneous, rare and not fully characterized group of diseases.

Objectives: Evaluation of ES patient’s clinical features and its evolution.

Methods: 47 patients with >20% BAL eosinophils or compatible histological features were included and its clinical files retrospectively analyzed.

Results: After excluding 19 patients that had other diseases and 9 because of lacking information, 19 patients were enrolled: 8 presented Chronic Eosinophilic Pneumonia (CEP), 6 Churg-Strauss Syndrome (CSS), 3 Acute Eosinophilic Pneumonia and 2 Allergic Bronchopulmonary Aspergillosis (ABPA). Mean age at symptoms onset 37±14.6 and at diagnosis 43±14.3 years. The majority was female (68.4%) and nonsmokers (76.5%). Peripheral eosinophilia was observed in 78.9% patients (1.7±1.32x10^9/L). Male gender, X-ray stage I or II, lymphocytic alveolitis in bronchoalveolar fluid, spontaneous regression, and active disease duration up to 2 years appear to be good prognostic factors.