Polish Pneumonology and Allergology

Pneumonologia i Alergologia Polska

Formerly **PNEUMONOLOGIA POLSKA**, edited since 1926

ABSTRACTS

8th International WASOG Conference on Diffuse Parenchymal Lung Diseases

June 2–4 2016, Gdańsk

The Journal is indexed in the following databases: Index Medicus/Medline, EMBASE, EBSCO, Scopus, Index Copernicus (124,2 2014). MNiSW (13 points 2014). Access http://jml2012.indexcopernicus.com/

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VIA MEDICA

THURSDAY, JUNE 2TH, 2016

07.00 am	**Registration**
08.15–08.30 am	**Welcome/Introductions**
SESSION 1. EDUCATION ON SARCOIDOSIS; NOT ONLY FOR PATIENTS… **(This session will be translated into Polish)** **Chairpersons:** *Marjolein Drent (Bennekom, Netherlands), Robert P. Baughman (Cincinnati, USA), Elyse E. Lower (Ohio, USA), Emer Joyce (Cleveland, USA), Agnieszka Staniewicz-Panasik (Gdansk, Poland)*	
08.30–08.50 am	**Pulmonary sarcoidosis** *Robert P. Baughman (Cincinnati, USA)*
08.50–09.10 am	**Update on cardiac sarcoidosis** *Emer Joyce (Cleveland, USA)*
09.10–09.30 am	**Ocular sarcoidosis. Overview** *Agnieszka Staniewicz-Panasik (Gdansk, Poland)*
09.30–09.50 am	**Fatigue syndrome** *Elyse E. Lower (Ohio, USA)*
09.50–10.10 am	**Vitamin D and calcium metabolism in sarcoidosis** *Robert P. Baughman (Cincinnati, USA)*
10.10–10.30 am	Coffee break
10.30–10.50 am	**Beyond corticosteroids. Alternative therapeutic options** *Marjolein Drent (Bennekom, Netherlands)*
10.50–11.00 am	**Presentation. Foundation for Sarcoidosis Research** *Ginger Spitzer, Executive Director of Foundation for Sarcoidosis Research*
11.00–11.45 am	**Question and Answer**
SESSION 2. AETIOLOGY, PATHOLOGY, AND DIAGNOSIS OF INTERSTITIAL LUNG DISORDERS (ILD) **Chairpersons:** *Katerina M. Antoniou (Crete, Greece), Ganesh Raghu (Washington, USA), Venerino Poletti (Forlì, Italy)*	
12.00–12.25 pm	**Update of classification of IIPs** *Katerina M. Antoniou (Crete, Greece)*
12.25–12.50 pm	**Demystifying the diagnosis of ILD. Is there a need for multidisciplinary clinical-radiologic-pathologic approach to diagnosis of ILD?** *Ganesh Raghu (Washington, USA)*
12.50–01.15 pm	**Transbronchial cryobiopsy in diffuse parenchymal lung diseases** *Venerino Poletti (Forlì, Italy)*
01.15–01.40 pm	**Serum and BAL biomarkers in ILD** *Jan C. Grutters (Nieuwegein, Netherlands)*
01.40–02.30 pm	**Meeting with the former President of Poland — Lech Wałęsa**
02.10–03.10 pm	Lunch
SESSION 3. COMORBIDITIES, CLINICS, REHABILITATION, AND LUNG TRANSPLANTATION IN ILD **Chairpersons:** *Sara Tomassetti (Forlì, Italy), Dominique Israel-Biet (Paris, France)*	
03.10–03.35 pm	**Pulmonary hypertension in ILD** *Katerina M. Antoniou (Crete, Greece)*
03.30–04.00 pm	**Genetic in ILD — from children to adults** *Bruno Crestani (Paris, France)*
04.00–05.00 pm	**Case presentation. A clinical round** *Sara Tomassetti (Forlì, Italy)*
05.00–05.25 pm	**Lung transplantation in ILD** *Dariusz Jastrzębski (Katowice, Poland)*
05.25–05.30 pm	**Summary and conclusions**
05.30 pm	**Opening ceremony**
05.30 pm	**Welcome** *Anna Dubaniewicz*
05.45 pm	**Welcome** *Marjolein Drent*
06.00 pm	**Leszek Możdżer's performance**
07.00 pm	Banquet

FRIDAY, JUNE 3RD, 2016 — INTERSTITIAL LUNG DISEASES/IPF	
SESSION 4. IDIOPATHIC PULMONARY FIBROSIS (IPF) **Chairpersons:** *Bruno Crestani (Paris, France), Athol Wells (London, UK), Antje Prasse (Hannover, Germany)*	
08.00–08.25 am	**Pathogenesis of IPF. What's new?** *Antje Prasse (Hannover, Germany)*
08.25–08.50 am	**Strategies to improve early diagnosis in IPF** *Athol Wells (London, UK)*
08.50–09.15 am	**Micro-CT research data on airways in IPF** *Wim Wuyts (Leuven, Belgium)*
09.15–09.40 am	**Staging of IPF** *Bruno Crestani (Paris, France)*
09.40–10.00 am	**Exacerbations of IPF** *Wojciech Piotrowski (Lodz, Poland)*
10.00–10.30 am	Coffee break
10.30–12.00 pm	**SATELLITE SYMPOSIUM ORGANISED BY BOEHRINGER INGELHEIM. CHALLENGES AND CURRENT TREATMENT APPROACHES IN IPF** **Chairperson:** *Luca Richeldi (Modena, Italy)*
10.30–10.35 am	**Welcome** *Luca Richeldi (Modena, Italy)*
10.35–10.50 am	**Lessons learned from recent IPF trials** *Ganesh Raghu (Washington, USA)*
10.50–11.10 am	**Real-life scenarios. Current treatment approaches in IPF** *Luca Richeldi (Modena, Italy)*
11.10–11.25 am	**The potential role of personalised medicine in future treatment of IPF** *Imre Noth (Chicago, USA)*
11.25–11.45 am	**Chronic disease management and palliative care in patients with IPF** *Michael Kreuter (Heidelberg, Germany)*
11.45–12.00 pm	**Panel discussion and closing remarks**
12.00–01.00 pm	Lunch
01.00–02.00 pm	**SATELLITE SYMPOSIUM ORGANISED BY ROCHE. NEW DIRECTIONS FOR IPF TREATMENT. CLINICAL TRIAL DATA AND PRACTICAL MANAGEMENT** **Chairperson:** *Jan Kuś (Warsaw, Poland)*
01.00 pm	**Welcome** *Jan Kuś (Warsaw, Poland)*
01.00–01.20 pm	**The evolution of pharmacological treatment of IPF. Learnings from clinical trial data** *Urlich Costabel (Essen, Germany)*
01.20–01.40 pm	**Pharmacological treatment of IPF. Recent insights** *Wim Wuyts (Leuven, Belgium)*
01.40–02.00 pm	**Long-term management of patients with IPF** *Vincent Cottin (Lyon, France)*
SESSION 5. RARE ILD **Chairpersons:** *Francesco Bonella (Essen, Germany), Daniel Culver (Cleveland, USA), Martin Petrek (Prague, Czech Republic)*	
02.00–02.20 pm	**Combined pulmonary fibrosis and emphysema. A clinical phenotype of IPF** *Francesco Bonella (Essen, Germany)*
02.20–02.40 pm	**Rare PPFE may occur also in your consulting room** *Ewa Jassem (Gdansk, Poland)*
02.40–03.00 pm	**Lymphangioleiomyomatosis and pulmonary Langerhans cell histiocytosis. Advances in diagnosis and therapy** *Elżbieta Radzikowska (Warsaw, Poland)*
03.00–03.20 pm	**Cutting-edge in the pathogenesis and treatments of pulmonary alveolar proteinosis** *Daniel Culver (Cleveland, USA)*
03.20–03.50 pm	Coffee break
SESSION 6. OTHER ILD **Chairpersons:** *Urlich Costabel (Essen, Germany), Arata Azuma (Tokyo, Japan), Agnieszka Jarzemska (Bydgoszcz, Poland)*	
03.50–04.10 pm	**Interstitial pneumonia with autoimmune features: relevance for clinicians** *Vincent Cottin (Lyon, France)*
04.10–04.30 pm	**Hypersensitivity pneumonitis** *Urlich Costabel (Essen, Germany)*
04.30–04.50 pm	**Drug-related ILD. New insights** *Marjolein Drent (Bennekom, Netherlands)*
04.50–05.10 pm	**Occupational ILD** *Arata Azuma (Tokyo, Japan)*

SESSION 7. POSTER SESSION **Chairpersons:** *Marjolein Drent (Bennekom, Netherlands), Elyse E. Lower (Ohio, USA), Robert P. Baughman (Cincinnati, USA), Urlich Costabel (Essen, Germany), Arata Azuma (Tokyo, Japan), Daniel Culver (Cleveland, USA), Anna Dubaniewicz (Gdansk, Poland), Hui-Ping Li (Shanghai, China)*	
05.10–06.00 pm	**Poster discussion**
06.00–06.10 pm	**Summary and conclusions**
08.00–12.00 am	**Gala dinner**

SATURDAY, JUNE 4TH, 2016 — SARCOIDOSIS

SESSION 8. AETIOLOGY, DIAGNOSIS AND FOLLOW-UP OF SARCOIDOSIS **Chairpersons:** *Elyse E. Lower (Ohio, USA), Robert P. Baughman (Cincinnati, USA), Anna Dubaniewicz (Gdansk, Poland)*	
08.00–08.25 am	**Progenitor cells in pulmonary sarcoidosis** *Tadeusz Płusa (Warsaw, Poland)*
08.25–08.50 am	**Immunogenetics** *Natalia Rivera (Stockholm, Sweden)*
08.50–09.15 am	**Microbiome in sarcoidosis** *Elżbieta Puścińska (Warsaw, Poland)*
09.15–09.35 am	**Sarcoidosis and tuberculosis: similarities and differences** *Anna Dubaniewicz (Gdansk, Poland)*
09.35–10.05 am	Coffee break
10.05–10.30 am	**Diagnosis: an update. Scintigraphy, HRCT, and PET/MRI/CT in sarcoidosis** *Dariusz Ziora (Katowice, Poland)*
10.30–10.55 am	**Diagnosis: an update. EBB, TBLB cTBNA, EBUS-TBNA or EUS-FNA?** *Artur Szlubowski (Zakopane, Poland)*
10.55–11.20 am	**Sarcoidosis vs. sarcoidal reaction — a role of cytopathological diagnosis** *Joanna Domagała-Kulawik (Warsaw, Poland)*
11.20–11.55 am	**Follow-up of sarcoidosis** *Robert P. Baughman (Cincinnati, USA)*
11.55–12.25 pm	**The presentation of best three posters**
12.25–01.25 pm	Lunch
SESSION 9. TREATMENT OF SARCOIDOSIS. CONVENTIONAL AND NEW TREATMENTS **Chairpersons:** *Dominique Israel-Biet (Paris, France), Robert P. Baughman (Cincinnati, USA)*	
01.25–01.50 pm	**Standard medical management of sarcoidosis** *Daniel Culver (Cleveland, USA)*
01.50–02.15 pm	**Benefits of rehabilitation in sarcoidosis** *Bert Strookappe (Bennekom, Netherlands)*
02.15–02.35pm	**A norel rehabilitation exercise for the patients with pulmonary fibrosi** *Hui-Ping Li (Shanghai, China)*
03.00–03.25 pm	**Acthar and other novel treatments of sarcoidosis** *Robert P. Baughman (Cincinnati, USA)*
03.25–03.55 pm	**Challenges in designing clinical trials. Sarcoidosis vs. pulmonary fibrosis** *Eric Meltzer (Durham, USA)*
03.25–03.55 pm	**Presentation. Foundation for Sarcoidosis Research** *Ginger Spitzer, Executive Director of Foundation for Sarcoidosis Research*
03.55–04.20 pm	**Summary of the Conference** *Daniel Culver (Cleveland, USA)*

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Contents

Abstracts

P1

Angiotensin-converting enzyme in serum: false positives? A clinical case

Joana Clemente Duarte; Margarida Afonso; António Jorge Ferreira
Coimbra Hospital and University Center, Coimbra, Portugal

Introduction: Angiotensin-converting enzyme in serum (ACES) is one of the key elements that increase the level of suspicion for the diagnosis of pulmonary sarcoidosis, a chronic systemic granulomatous disease of unknown origin, probably secondary to an abnormal immune response. Its association with lymphomas are described, including the description of „sarcoidosis-lymphoma syndrome" (SLS). There are few cases of SLS and the nature of this relationship is poorly characterized.
Case report: A 47 year old woman with no medical history was being followed since 2012 for a pulmonary nodule that was stable, without lymphadenopathy associated. After three years, after a pneumonic process, a CT scan showed multiple mediastinal, supra-clavicular and lateral-cervical lymph nodes. Blood sample analysis revealed a high value of ACES. Autoimmunity, protein profile, immunoglobulins and determination of calcium in the urine were normal. The patient was submitted to positron emission tomography, which revealed hypermetabolic lateral-cervical node and hilar involvement and underwent surgical removal of a supraclavicular node. The clinical pathology showed "gray area" lymphoma. Chemotherapy protocol was initiated and imaging control showed no lateral-cervical, mediastinal and hilar lymphadenopathy. She remains asymptomatic.
Conclusions: Although this is not a typical case of "sarcoidosis-lymphoma syndrome" it highlights how increased levels of ACES may occur in these two diseases and lymphoma's image can provide similar results as in pulmonary sarcoidosis. The clinicians should be aware that these pathologies can coexist and in some cases is still essential to achieve a correct histopathological diagnosis with implications in treatment, evolution and disease prognosis.

P2

Sarcoidosis and veinous thromboembolic disease: about five observations

Frederic Riviere; Pierre Louis Conan; Herve Le Floch; Olivier Bylicki; Wanda Gaspard; France Charton; Jacques Margery
Percy military hospital, Clamart, France

Introduction: Sarcoidosis, as chronic inflammatory disorders, is an unknow risk factor for veinous thromboembolic disease (VTED) 1, 2. Aim. For the first time, we are describing five observations of VTED associated with sarcoidosis.
Material and methods: Retrospective analysis of the epidemiological, clinical, radiological and biological data of patients who experienced a VTED (pulmonary embolism [PE] or deep vein thrombosis [DVT]) associated to histologically proved Sarcoidosis, previously or synchronously diagnosed.
Results: 4 men and one woman, all of them Caucasians with a median age of 49 years old [46; 67] have showed 2 PE, 2 DVT and one combinaison of both. Two patients had a previous history of DVT. It was the first sign of the sarcoidosis for 4 cases out of 5. Three mediastinal- pulmonary sarcoidosis were stage 1 or 2, 1 had pulmonary and cardiac involvement and one neurosarcoidosis. They were considered to be relatively inactive: lymphocytes 1128/mm 3 [918; 1470], gammaglobulineaemia 9.2 g/L [8.7; 10.5], calciuria 2.21 mmol/day [2.11; 6.23] and no volume restriction nor airflow obstruction with a median FEV1/FVC of 78. 81%. Three patients presented another VTED when anticoagulants were discontinued and all of them were evolving toward a chronical sarcoidosis without refractory cases. They weren't showing another VTED risk factor3 and thrombophilia search was negative. Activity markers like chitotriosidase or neopterin weren't performed.
Conclusions: Even if relative risk is low4, sarcoidosis might be considered as a risk factor for VTED.

P3

Langerhans cell histiocytosis: variability in pulmonary involvement

Joao Silva; Tito Abrantes; Marta Sousa; Vitor Melo; Antonio Reis; Ana Campos; Simoes Torres
Centro Hospitalar Tondela-Viseu, Viseu, Portugal

Introduction: Langerhans cell histiocytosis (LCH) or histiocytosis X, is a rare disorder characterized by an abnormal increase in histiocyte cells, components of the immune system. Signs of LCH depend on the extent and location of the disease. A male predominance is observed. Clinical cases The author presents two clinical cases with distinct pulmonary involvement.
Case 1: 18 year-old male, smoker, with chronic dry cough and acute chest pain. Computed tomography (CT) of the chest showed exuberant pulmonary cysts, diffusely distributed with a predominance in the lung apices, unequal size and bizarre shapes. Bronchofibroscopy with bronchoalveolar lavage was negative to CD1a cells, but lung biopsy established the diagnosis.
Case 2: 19 year-old male, non-smoker, with sudden dyspnea and pleuritic chest pain. Chest x-ray pointed out left spontaneous pneumothorax, which wasn't solved with thoracic drainage. He has undergone cirurgical pleurodesis with blebs resection whose pathological report was compatible with LCH. Thorax CT identified only small cystic lesions in the right pulmonary apex.
Conclusions: In adults with LCH, the pulmonary system is the most frequently involved organ and pulmonary lesions may be the only manifestation. The clinical spectrum is broad. Image studies can reveal from diffuse cystic lesions, nodular infiltrates, pleural effusion and pneumothorax to minimal changes of the parenchyma.

P4

Sarcoidosis incidence and prevalence: a nationwide register-based assessment in Sweden

Elizabeth V. Arkema; Johan Grunewald; Susanna Kullberg; Anders Eklund; Johan Askling
Karolinska Institutet, Stockholm, Sweden

Aim: To estimate the contemporary incidence and prevalence of sarcoidosis using Swedish national population-based register data.
Material and methods: Adults with any visit listing an ICD code for sarcoidosis were identified from the National Patient Register (hospitalizations 1964−2013 and outpatient care 2001−2013). Demographic and medication dispensing data were retrieved from national registers. We estimated the prevalence of sarcoidosis in 2013 overall and by county of residence. Incident sarcoidosis 2003−2012 was estimated overall, by sex, age, education level and year of diagnosis. Definitions of incident and prevalent cases were varied to test the robustness of our results.
Results: Over 16, 000 people had a history of sarcoidosis in 2013. When defined as 2 sarcoidosis-coded visits, the prevalence was 160/100, 000. Using different definitions, the prevalence ranged from 152 (requiring a specialist visit) to 215/100,000 (only 1 visit required). The highest prevalence was observed in northern less population-dense counties. There were over 10, 000 incident cases between 2003 and 2012. The incidence was 11.5/100,000 per year and varied by −10% to +30% depending on case definition. The incidence was highest in males 30−50 years old and in females 50−60 years old but did not differ by education level and was stable over time.
Conclusions: This study represents the largest epidemiological investigation of sarcoidosis using population-based individual-level data. Sarcoidosis occurrence was stable over time but varied substantially by geographical region, age and sex. The age at onset was 10 years younger in males than in females.

P5

Antisynthetase syndrome: case series of a rare condition

Joana Clemente Duarte; Margarida Afonso; Sara Freitas; Tiago Alfaro; Carlos Robalo Cordeiro
Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Introduction: The antisynthetase syndrome (ASS) is a rare auto-immune disorder characterized by a variable association between antisynthetase antibodies and interstitial lung disease, myositis, arthritis, fever, Raynaud's phenomenon and mechanic's hands. Pulmonary involvement, when present, is the main prognosis determinant. AIM: Our objective was to characterize the ASS population under the care of an interstitial lung disease (ILD) clinic in a university hospital in Portugal.
Material and methods: We performed a retrospective analysis of all ASS cases attending our clinic between 2006 and 2015. Patients were identified through a search of the clinical database. We collected data on complaints, lung function, imaging, laboratory, treatment and prognosis.
Results: A total of six patients (five female and one male) were included, with a median age of 58 years. All were white, with a median age at diagnosis of 50 years. Most presented for systemic complaints, including asthenia and myalgia. One had skin lesions. None of the subjects had neurological or cardiac involvement. Lung function was normal in three, while the others had mild restriction with low diffusion capacity. On chest imaging, half showed predominant ground glass infiltrates, and half had predominant fibrotic changes. All received chronic steroids, with concomitant azathioprine in most. Upon dose reduction, four patients had symptom relapse. No deaths were registered during a median follow-up of 48 months.
Conclusions: In our patient population, ASS affected mostly white middle aged females. There was a good clinical response to treatment, but a high rate of relapse upon reduction of steroids.

P6

Fatal arrhythmic events in patients with cardiac sarcoidosis; retrospective cohort analysis of Japan

Kengo Kusano[1]; Yusuke Hattori[1]; Kohei Ishibashi[1]; Teruo Noguchi[1]; Toshihisa Anzai[1]; Satoshi Yasuda[1]; Hisao Ogawa[1]; Fumio Terasaki[2]; Mitsuaki Isobe[3]
[1]*National Cerebral and Cardiovascular Center, Suita, Japan*; [2]*Osaka Medical College, Takatsuki, Japan*; [3]*Tokyo Medical and Dental University, Tokyo, Japan*

Introduction/Aim: Sarcoidosis is a systemic non-caseating granulomatous disease of unknown etiology. Cardiac involvement (cardiac sarcoidosis, CS) has been reported to be an important prognostic factor and corticosteroid is usually used to prevent deterioration of cardiac function and fatal arrhythmia. We evaluated the long-term prognosis in patients with CS from Japanese Multi-Center retrospective cohort analysis.
Material and methods: Total of 746 Japanese patients from 56 hospitals who diagnosed CS were examined. Patients who unsatisfied the criteria for cardiac sarcoidosis proposed by the Japanese Society of Sarcoidosis and Other Granulomatous disease (JSSOG) modified in 2006 were excluded, and 438 patients (135 males, mean age of 59.7 ± 12.1 years old, and mean follow-up period of 5.6 ± 4.7 years) were analyzed. The relationship of cardiac function (left ventricular ejection fraction: LVEF) and later adverse events were evaluated.
Results: Corticosteroid (mean dose of 7. 2 mg) was prescribed in 382 of CS patients (87.2%) and total 97 adverse events (22.1%) were observed (all-cause death in 52 and appropriate implantable cardioverter defibrillator (ICD) discharge in 59 patients). Kaplan-Meier analysis revealed that the fatal arrhythmic events (sudden death and appropriate ICD discharge) were more observed in moderate or low LVEF by initial echocardiography (log-rank $p < 0.02$). This observation was also observed in patients without prior ventricular arrhythmia.
Conclusions: This large scale Japanese multi- center data showed that initial LVEF was a strong predictor of fatal arrhythmic events in CS, therefore ICD implantation as a primary prevention would be needed even after starting corticosteroid in patients with decreased cardiac function.

P7

Reactivation inflammation impacts on the ventricular arrhythmia for the patients with cardiac sarcoidosis

Kenzaburo Nakajima[1]; Ikutao Nakajima[2]; Kohei Ishibashi[1]; Tsukasa Kamakura[1]; Mitsuru Wada[1]; Yuko Inoue[1]; Koji Miyamoto[1]; Takashi Noda[1]; Takeshi Alba[1]; Shiro Kamakura[3]; Kengo Kusano[1]
[1]*National Cardiovascular Center, Osaka, Japan*; [2]*St. Marianna medical university, Kawasaki, Japan*; [3]*National Cardiovascular Centerati, Osaka, Japan*

Introduction: Cardiac sarcoidosis (CS) sometimes developed lethal ventricular arrhythmia (VA). The relationship between reactivation of inflammation after steroid therapy and VA in patients with CS is still unclear.
Material and methods: Data on 121 CS patients with reduced LVEF and steroid therapy were enrolled. The inflammation status was evaluated by cardiac-positron emission tomography and Gallium-shintigraphy every 6 months. We evaluated relationships with reactivation of inflammation and VA events.
Results: During a median follow-up of 1418 days, 25% of patients showed reactivation of inflammation. VA events were significantly higher in patients with reactivation than others ($p < 0.01$). Interestingly, the type of VA manifestation was different and ventricular fibrillation was more common in patients with inflammation reactivation.
Conclusions: Reactivation of inflammation in patients with CS is a highly risk of ventricular arrhythmia.

P8

Clinical value of HRCT scores in newly diagnosed pulmonary sarcoidosis

Regina Aleksoniene[1]; Ingrida Zeleckiene[2]; Mindaugas Mataciunas[2]; Roma Puronaite[3]; Edvardas Danila[1]
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Aim: To evaluate the significance of high-resolution computed tomography (HRCT) scores and impairment of pulmonary function in different radiologic stages of sarcoidosis patients.
Material and methods: 80 patients with newly diagnosed sarcoidosis (41 F, 39 M; age 39 ± 9 yrs) were included. Patients underwent HRCT scanning and pulmonary function tests (PFT). HRCT scores and PFT values were analyzed. 15 (19%) of patients had stage 1, 59 (74%) — stage 2, and 6 (7%) — stage 3.
Results: Micronodules (77.5%), macronodules (53.8%) and linear opacities (45%) were most frequently met patterns in HRCT of patients with pulmonary sarcoidosis. Only the number of micronodules was significantly higher ($p = 0.004$) in stage III compared with other stages.
Statistically significant differences between FVC ($p = 0.040$), FEV_1 ($p = 0.017$), TLC ($p = 0.049$), VC ($p = 0.015$), DLCO ($p = 0.012$) were found in different radiographic stages of sarcoidosis. PFT values were decreased in stage 3 compared with stage 1 patients.
Significant correlations between consolidation score on HRCT and FVC ($r = -0.227$, $p = 0.043$), FEV_1 ($r = -0.299$, $p = 0.007$), FEV_1/FVC ($r = -0.245$, $p = 0.029$) as well as correlation between ground glass opacities score and DLco ($r = -0.267$, $p = 0.017$) were established. We did not found significant correlations between micronodules or macronodules score and PFT indices.
Conclusions: A strong interface between radiologic changes and pulmonary function impairment exists in sarcoidosis patients. Consolidation and groud glass opacities HRCT scores, but not micronodules or macronodules scores correlated negatively with pulmonary function indices. However, overall clinical value of HRCT scores was not very high.

P9

Telomere shortening in type II pneumocytes relates to idiopathic pulmonary fibrosis

Aernoud van Batenburg[1]; Reinier Snetselaar[1]; Roel Goldschmeding[2]; Tri Nguyen[2]; Matthijs van Oosterhout[1]; Karin Kazemier[1]; Jan Grutters[1]; Coline van Moorsel[1]
[1]*St. Antonius Hospital Nieuwegein, Nieuwegein, The Netherlands*; [2]*UMC Utrecht, Utrecht, The Netherlands*

Introduction: Idiopathic Pulmonary Fibrosis (IPF) is a devastating disease with a median post-diagnostic survival of approximately 4 years. Recently it has been discovered that telomere length (TL) maintenance plays a key role in IPF. In patients with different types of pulmonary fibrosis, shortening of leukocyte telomere length was found, especially in telomerase mutation-related disease. In this pilot study, we measured TL in the regenerating cells of lung alveoli, so-called type II pneumocytes. These cells are considered to be the crucial cell type in IPF pathogenesis.
Material and methods: Tissue slides of sporadic IPF patients and controls were stained using telomere Fluorescence In Situ Hybridization (FISH). Type II cells were identified using the proSPC antibody from Merck Millipore (ab3786, Rabbit 1: 300). Subsequently, quantification of telomere fluorescence was measured per cell type, discriminating between type II and all the surrounding cells combined. These were classified as non-type II cells.
Results: Type II pneumocyte TL was significantly shorter in fibrotic areas compared to non-fibrotic areas or control tissue (p-values of 0.004 and 0.0023 respectively). In non-type II pneumocytes TL did not differ between fibrotic and non-fibrotic areas. Furthermore, within controls no differences were observed between type II cells and non-type II cells.
Conclusions: In conclusion, the co-localization of short telomeres in type II cells and the pathophysiology of IPF suggests that telomere shortening plays a critical role in the fibrotic remodeling of lung tissue in this disease.

P10

Airway-centered interstitial fibrosis — rare interstitial lung disease: 2 case reports

Vitor Melo; Marta Sousa; Joao Silva; Tito Abrantes; Ana Campos; Simoes Torres
CHTV, Viseu, Portugal

Airway-centered interstitial fibrosis (ACIF) is a new and rare interstitial lung disease (ILD) of unknown cause characterized by chronic cough and progressive dyspnea; history of inhaled exposure; propensity for women; occurrence in middle age; progressive peribronchiolar distribution of interstitial inflammation and fibrosis with bronchiolar metaplasia distinct from any form of ILD or idiopathic interstitial pneumonia (IIP). The majority of patients are non-responsive to corticotherapy with poor prognosis. We describe 2 female patients, 65 and 66 years old, presenting with chronic dry cough and progressive dyspnea. Both non-smokers, farmers and with a history of inhaled exposition to birds. Chest radiographs revealed diffuse reticulonodular infiltrates and bronchial walls thickening. ChestCT showed reticular fibrosis and ground glass infiltrates across the pulmonary fields in one and sparing the upper on the other. Bronchoalveolar lavage showed a small increase in lymphocytes/neutrophils in one, the ratio CD4/CD8 < 1 in both. The diagnosis was made by surgical biopsy revealing pericentrilobular lesions and linfoplasmocitary infiltrate compatible with ACIF. Both patients began systemic corticotherapy for 12 months. In one case the disease progressed with worsening symptoms, pulmonary functional tests and CT imaging even after starting combined imunosuppression with Azathioprine. The other patient improved with less symptoms and decreased area of disease on the CT image. These 2 cases presented similar clinical, radiological and pathological features as most of the few cases reported in literature. Our patients were non-smokers but were exposed to birds. The evolution varied with one improving after therapy and the other progressing even after combined imunosupression.

P11

Clarithromycin decreases IL-6 concentration in serum and BAL fluid in patients with cryptogenic organising pneumonia

Elżbieta Radzikowska; Adriana Roży; Paulina Jaguś; Małgorzata Polubiec-Kownacka; Joanna Chorostowska-Wynimko; Kazimierz Roszkowski-Śliż; Elżbieta Wiatr
National Tuberculosis and Lung Diseases Research Institute, Warsaw, Poland

Introduction: Inflammatory cytokines are involved in the development of cryptogenic organising pneumonia (COP). It was shown, that macrolides inhibited the cytokines production in alveolar macrophages of COP patients. Objectives: Assessment of interleukin 1β (IL-1β), IL-6, IL-8, and transforming growth factor β (TGF-β) concentrations in serum, and in BAL fluid (BAL-f) in COP patients treated with clarithromycin.
Material and methods: 26 patients with biopsy proven COP (18 women and 8 men), in a mean age of 56. 46 ± 8. 83 years were enrolled into the study. Complete response was achieved in 22 patients and four patients did not respond to treatment. ELISA method was used to measure the serum and BAL-f concentrations of IL-1β, IL-6, IL-8, and TGF-β.
Results: Before treatment the serum IL-1β1, IL-6, IL-8, and TGF-β1 concentrations were similar in responders and non- responders. Significant decrease of IL-6 [8. 98 ± 13. 26 pg/mL vs. 3. 1 ± 6.95 pg/mL; p = 0.005)], IL-8 [20.14 ± 25.72 pg/mL vs. 10.14 ± 6.8 pg/mL; p = 0.007], and TGF- β1 [37.89 ± 12.49 ng/mL vs. 26.49 ± 12.45 ng/mL; p = 0.001] serum concentration, and IL-6 concentration [30.56 ± 56.78 pg/mL vs. 4. 53 ± 5.84 pg/mL; p = 0.036)] in BAL-f was noticed after clarithromycin (CAM) treatment. Clarithromycin treatment resulted in significantly lower mean value of IL-6 in serum in responders than in non- responders.
Conclusions: Response to clarithromycin treatment was associated with decrease of IL-6, IL-8, and TGF-β1serum concentrations, and IL-6 concentration in BAL-f.

P12

Results of clarithromycin treatment in patients with cryptogenic organising pneumonia

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Introduction: Despite that corticosteroids have been established as the standard treatment of cryptogenic organising pneumonia (COP), other drugs such as clarithromycin (CAM), are also proved as effective. The goal of this study was to present the results of CAM versus prednisone (PRE) treatment in patients with biopsy-proven COP.
Material and methods: From 1999 to 2014, 44 patients were treated with CAM (500 mg twice daily orally, for 3 months) and 32 with PRE (mean initial dose of 0. 73 ± 0.24 mg/kg/d for a mean of 8. 68 ± 4.08 months).
Results: The clinical presentation, laboratory, and radiological findings did not differ significantly between patients treated with CAM and PRE, with the exception of a higher frequency of sweats, ground glass opacities, and migratory pattern of lesions, and fewer cases of FEV_1 of < 80% and hypoxemia among patients treated with CAM than with PRE. A complete response was achieved in 39 (93%) patients treated with CAM and in all treated with PRE. Relapse was noted more frequently in patients treated with PRE than with CAM (53% vs. 7%; p < 0.0001). Adverse events were noticed in 53% of patients treated with PRE and 2% treated with CAM. FVC > 78% of predicted with sensitivity — 79% and specificity-80%, and FEV_1 > 63% of predicted with sensitivity −97% and specificity — 60% identified patients who were good candidates for CAM treatment.
Conclusions: CAM can be used as an alternative treatment of COP, but in patients with pulmonary function parameters within the normal limits. This therapy is shorter, better tolerated, and associated with fewer adverse events and relapses than PRE.

P13

Chronic hypersensitivity pneumonitis: implications of usual interstitial pneumonia pattern in disease severity

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Introduction: Chronic hypersensitivity pneumonitis (cHP) has a wide variable clinical presentation and evolution, and there is a subGroup of patients with usual interstitial pneumonia pattern (cHP/UIP) which information is scarce.
Aim: Characterization of patients with cHP, comparing cHP/UIP with other cHP.
Material and methods: Analysis of cPH patients with disease behaviour stratified according with ATS/ERS classification for IIP. HRCT fibrosis score was used for quantification of disease extent in cHP/UIP patients.
Results: Included 86 patients with mean (± SD) age of 61.5 (± 14) years, 66.3% (n = 57) female. Most (84.5%) were never smokers. Almost half (48.8%) had cHP/UIP, with an average fibrosis score of 8.4 (± 2.8). Patients with cHP/UIP had significantly lower DLCO [44.5 (± 18.5)% vs. 57 (± 21.5)%, p = 0.015] and lymphocytosis [27.1 (± 22.6)% vs. 45 (± 19.6)%, p = 0.001]. Regarding disease behaviour in cHP/UIP, 9.1% had stable disease (vs. 31.8% in other cHP), 45.5% had progressive, irreversible disease with potential for stabilization (vs. 38.6% in other cHP) and 45.5% had progressive, irreversible disease despite therapy (vs. 29.5% in other cHP). Therapeutic approach included steroids and immunosuppressants in 62.2%, only steroids in 21.6% and 16.2% didn't make any anti-inflammatory therapy, without relevant differences between Group s. Fourteen patients were referred for lung transplantation, 8 with cHP/UIP. During the follow-up, 24.4% died, with a median survival significantly lower in cHIP/UIP (106 vs. 154 months, p = 0.023).
Conclusions: cHP showed a wide variability in disease severity, standing out the Group with cHP/UIP in whom an unfavourable evolution occurred despite therapy. At diagnosis, this subGroup presented a greater functional severity and may require higher monitoring and therapeutic approach.

P14

Addition of EBUS-TBNA to bronchoscopic biopsies in the diagnosis of sarcoidosis

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Introduction: While the utility of endobronchial ultrasound-guided transbronchial needle aspiration (EBUS- TBNA) has been reported in providing sarcoidosis diagnosis, its performance in compunction with other bronchoscopic sampling methods remains to be clarified. Aims to evaluation the sensitivity of different diagnosis modalities (EBUS-TBNA, transbronchial biopsy-TBB, endobronchial biopsy-EBB) performed during the same endoscopic procedure for confirming sarcoidosis.
Material and methods: We retrospectively reviewed the data of 64 patients (20 men, 44 women), Ø 46 year old (range 31–69) in whom the suspected initial diagnosis of sarcoidosis had been supported by confirmation of granulomas. The diagnostic yield of the three bronchoscope sampling (EBUS-TBNA, TBB, EBB) between January 2012 and December 2015 in our Institute.
Results: According to radiologic CXR stages were: stage I 47%, stage II 34%, stage III 19%. Bronchoscopy with EBB was performed in 39/64, TBB in 55/64 and EBUS-TBNA of mediastinal and hilar lymph nodes 49/64. Confirmation of granulomas was achieved in 39% of EBB (regardless of macroscopic appearance), 80% of TBB and 66% of EBUS-TBNA. According to radiological stages (I vs. II) the diagnostic yield was: EBB 40% vs. 42%, TBB 30% vs. 92%, EBUS-TBNA 82% vs. 71%. There were 4 post-interventional pneumothoraxes.
Conclusions: EBUS-TBNA with combined samplings should be considered as a first diagnostic procedure in sarcoidosis stage I, but TBB seems to remain the better option for stage II disease.

P15

Clinical features of usual intertitial pneumonia — correlation between idiopathic pulmonary fibrosis and connective tissue disease associated with usual interstitial pneumonia

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Introduction: The study objective is to compare the clinical feature of IPF and UIP-CTD to address the clues for predicting the underlying situation.
Material and methods: A retrospective analysis of HRCT diagnosed UIP patients between 2004–2014 included demographic features, smoking habits, symptoms and signs, serology, pulmonary function and dominant radiology findings and selected treatment were compared between of IPF and UIP-CTD Group s.
Results: The total of 103 UIP patients (62 IPF and 41 UIP-CTD) were analysed. Most in the CTD patients had rheumatoid arthritis (n = 21), systemic sclerosis (n = 11), Sjögren syndrome (n = 8), systemic lupus erythematosus (n = 2), and polymyositis (n = 1). The IPF Group included 65% of males, while females made 60% of the UIP- CTD Group. The patients mean age was significantly different in the IPF and UIP-CTD Group –65.79 ± 10.15 vs. 60.0 ± 12.98. Smokers made 59% and 27% of the IPF and UIP-CTD Group respectively. IPF Group had severer and longer lasting symptoms, dyspnoea and cough, as well as the mean mMRC dyspnoea score. Clubbing was more frequently registered in IPF patents (39% vs. 19%). Autoantibodies were positive in 35% of IPF patients (RF, ANA). In IPF Group had lower mean DLco, TLC and RV. The only radiological findings were significantly different in the two Group s, presented with honeycomb lungs in 87% and 45% of the IPF and UIP-CTD Group respectively. Immune suppressive treatment was more commonly administered in the UIP-CTD Group (81% vs. 52%).
Conclusions: In the patients with UIP pattern in HRCT, IPF was most frequently diagnosed in older smoking males with clubbing. Autoantibodies are of no use in differentiating between IPF and CTD patients. IPF patients might be diagnosed lately since those patients mostly have a longer history of symptoms, reduced lung volumes and diffusion capacity, and more common honeycombing in HRCT. Typical extrapulmonary symptoms could alert physicians to investigate CTD or in CTD patients pulmonary symptoms may lead to investigate pulmonary involvement.

P16

The concept of organizing pneumonia — as a pathologic term

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Introduction: Organizing pneumonia (OP) is a histopathological term describing patchy filling of the lung alveoli and respratory bronchioles by loose plugs of granulation tissue. OP term may describe mainly OP disease or may be seen in association with another disease such as malignancy, infections. In present study, we investigated the clinical reflection of lesions defined as OP histopathologically.
Material and methods: Between 2011–2015, all lung pathology reports including the term'organizing penumonia'were included. All patients'; medical files were examined. OP were classified as reactive OP (adjacent to another predominant disease) and OP disease. The demographic, radiological and clinical findings were recorded. Radiological findings were Group ed as typical, focal and infiltrative.
Results: Among 6352 lung pathology reports, 138 reports were included. Thirty-four (25%) were female and the mean age was 54 ± 14. Six of the biopsies were transbronchial biopsy materials. 75 (54%) of the biopsies were right-lung sided. 84 (61%) of the patients were classified as reactive OP, 54 (39%) as OP disease. The most frequent reactive process was seen with malignancy (66%), mostly squamous cell carcinoma. The other diseases were bronchiectasis, lung abscess, tuberculosis, cyst hydatic, hypersensitivity pneumonia, bullae, etc. Forty-eight (89%) patients with OP disease, were diagnosed as COP. Radiological findngs were consistent with focal OP mostly (n = 34,63%). In PET-CT scans FDG uptake was high in focal forms.

Conclusions: OP, as a histopathologic term, is included in pathology reports describing reactive processes. Cryptogenic OP is more frequent than secondary forms. Radiologicallly focal OP is more often than the other forms.

P17

The clinical course of organizing pneumonia

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Introduction: The clinical presentation and clinical course of OP is quite variable. In present study, we investigated the clinical, radiological findings of OP patients and investigated the clinical courses.
Material and methods: The present study is a retrospective cohort study, conducted in a chest disease teaching hospital between 2005−2015. Patients diagnosed and followed as OP in our hospital were included. The clinical courses were categorized as progressive, relapse, non-relapse and the predictive factors for relapse were investigated.
Results: 19 (41%) were male and The mean age was 54 ± 14. The most common presenting symptoms were shortness of breath and cough. Twenty-five patients had no additional diseaes, 14 had never smoked. Radiological classification was consistent with typical OP in 31 (67%), focal in 7 and infiltrative in 8. The median follow-up duration was 18 (1−108) months. For treatment, steroid were used. Five patients had required intensive care unit follow-up due to respiratory failure and 3 of whom died within 2 months. Of the other 43 patients', 18 (39%) had at least one relapse. Median relapse duration was 7 months and 16 of them were re-treated with steroids. Two patients were decided to be treated life-long low-dose steroids for recurrent relapses. When the baseline parameters were investigated for relapse, demographics, smoking status, additional diseases, spirometric values, serum albumin, LDH values, cryptogenic or secondary forms, leucocyte, SaO_2 did not increase relapse risk. Lower levels of baseline hemoglobin ($p = 0.043$) and higher levels of bronchoalveolar lavage eosinophils ($p = 0.043$) increased relapse risk.
Conclusions: OP is mostly benign whereas rapidly-progressive courses may be seen. Lower baseline levels of hemoglobin and higher BAL eosinophils may predict relapsing forms.

P18

Novel reagents for prognosis follow-up on sarcoidosis

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Introduction: Sarcoidosis is a disease, which may be characterized by alveolitis and granuloma formation and result in fibrosis, and in the case of which spontaneous remissions may be observed as well. A test that, at the time of diagnosis, provides information about prognosis and enables the anticipation of the disease's likelihood of becoming chronic would be expedient. Mean platelet volume (MPV) and neutrophil-to-lymphocyte ratio (N/L) or Trombocyte-to-MPV ratio, which are readily and practically detectable currently, are such reagents, the prognostic significance of which have been proved for numerous inflammatory diseases. Objective: The objective of this study is to establish the significance of inflammatory reagent values at the time of initial hospital application of patients with sarcoidosis for the anticipation of the disease's likelihood of becoming chronic.
Material and methods: Some 142 of 384 cases followed up at our clinic, records for which have been maintained regularly for minimum two years from the relevant date of diagnosis, for which medical treatment has not been applied, and which are not at phase 0, were included to the study universe. The files of the patients were examined, and demographic details, relevant disease phase, mortality, follow-up period and inflammatory reagents (ACE, CRP, neutrophil, lymphocyte, trombocyte, MPV, N-to-L ratio, absolute N-to-L ratio and Trombocyte-to-MPV ratio) of the same were recorded. The cases in the study universe were Group ed into two with respect to the respective radiological healing progress; Group 1: The patients, in whose radiological findings no change was detected at the end of the two year period (chronic Group); Group 2: The patients, in whose radiological findings total remission was observed at the end of the two year period (remission Group).
Results: Group 1 was comprised of 86 cases (Median age: 47, F/M: 67/19), while Group 2 was comprised of 56 cases (Median age: 42, F/M: 45/11). The N-to-L ratio was detected to be 2.80 + −1.32 for Group 1 and 2. 45 + −1.62 for Group 2. It was observed that the LEN, N-to-L ratio and absolute N-to-L ratio of Group 2 was significantly lower than those of Group 1 ($p < 0.05$). Any significant difference was not established between the cases in Group 1 and those in Group 2 in terms of the other parameters ($p > 0.05$). The average follow-up period was established to be 65 months for Group 2 and 84 months for Group 1. The follow-up period for the patients in Group 1 was significantly longer than that for the patients in Group 2 ($p = 0.016$).
Conclusions: High values measured for the said reagents at the time of diagnosis in sarcoidosis cases can be considered as indicators suggesting delay in the remission of the patient, and can be used for the close follow-up of the patients.

P19

The function and importance of inflammatory reagents in researches on extrapulmanory involvement in sarcoidosis

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Introduction: Sarcoidosis may involve multiple organs or systems, among which lungs happen to be the most frequently involved organs. Researches on extrapulmonary involvement are of crucial importance for the monitoring and treatment of the patient. While, under treatment, the pulmonary lesions heal, the activity may continue in extrapulmonary organs, and the disease may even progress. Certain serum reagents (IL-18, sIL2R) come to the fore for their ability to indicate extrapulmonary involvement. The number of publications studying the use of Neutrophil-to-/lymphocyte (NLR) and Platelet-to-lymphocyte ratios, which are easily accessible at affordable costs, for the purpose of prognosis determination as a part of differential diagnosis has been increasing recently. Objective: The objective of this study is to research on the significance of the inflammatory reagent values of the sarcoidosis patients at the time of the initial hospital applications thereof in the determination of the extrapulmonary involvement.
Material and methods: 142 cases, which are followed up at our clinic and the extrapulmonary involvement of which have been studied, were included to the study universe. The files of the patients were examined, and the demographic details, disease phase, extrapulmonary involvement presence and inflammatory reagents (ACE, neutrophil, lymphocyte, trombocyte, MPV, NLR, NLR absolute, Trombocyte-to-MPV ratio) of the same were recorded. The cases were Group ed into two with respect to the extrapulmonary involvement presence;Group 1: no extrapulmonary involvement presence, Group 2: extrapulmonary involvement presence.
Results: Group 1 was comprised of 98 patients (Average of age: 44 ± 12, F/M: 77/21), while Group 2 was comprised of 44 patients (Average of age: 44 ± 11, F/M: 36/8);and while there were 103 patients at phase 1, there were 33 at phase 2 and 6 at phase 3. While Group 1 included 70 phase 1, 23 phase 2 and 5 phase 3 cases; Group 2 included 31 phase 1, 12 phase 2 and 1 phase 3 patients. The extapulmonary involvements observed in 44 patients were 3 eye involvements, 13 skin involvements and 3 heart involvements, respectively. The number of cases with double organ involvement was 12, while the number of triple organ involvement was 1. Any significant difference was not found amongst the cases in terms of ACE, neutrophil, lymphocyte, trombocyte, MPV, NLR, NLR absolute, Trombocyte-to-MPV ratio, being the reagents studied ($p > 0.05$).
Conclusions: The outcomes of the study reveal that ACE, neutrophil, lymphocyte, trombocyte, MPV, NLR, NLR absolute, Trombocyte-to-MPV ratio, being the inflammatory reagents studied, are not indicative in respect to the researches on the extrapulmonary organ involvement.

P20

Thyroid sarcoidosis: a case report

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Introduction: Sarcoidosis is a multisystemic disease with unknown etiology and it is characterized by non-caseating granuloma. Thyroid involvement of sarcoidosis is very rare and was first identified in 1938. The frequency of thyroid involvement in sarcoidosis cases was reported to be 4% based on

autopsy studies. In this report, we aimed to describe a rare case of thyroid involvement in sarcoidosis under the light of current literature.
Case: 54-years old female patient referred with the complaint of cough. Skin examination showed nodular lesions on the exterior skin of both arms. Hemogram and biochemistry parameters were within normal range. Lung X-ray showed bilateral hilar lymphadenopathy (BHL), fullness in right paratracheal region and density increase in right paracardiac region. Thorax CT showed bilateral nodules in thyroid glands, multiple mediastinal LAPs, BHL and bronchiectasis in the right middle lobe. In FOB, extensive millimetric mucosal nodules were detected in the bronchial mucosa of the intermediate and middle lobes. THNAB was initially performed from the main carina towards subcranial LAP, and then biopsy samples were obtained from the nodules by a forceps. Pathological diagnosis was reported as granulomatous inflammation with small necrotic foci. Skin biopsy revealed tuberculoid granuloma with central necrotic foci. Due to the thyroid lesion seen on thorax CT, left thyroidectomy and mediastinoscopy of the mediastinal lymph nodes were performed. Pathological examination of both tissues confirmed granulomatous inflammation with focal necrosis. Advanced investigations were performed for differential diagnosis. ACE was 47U/L, PPD was negative and PFT was found to be within normal range. Ophthalmology and cardiology consultations were normal. Sputum ARB was negative. The patient has been followed-up with these findings. PPD and ARB were still negative, and the skin lesions as well as lung findings spontaneously resolved after the follow-up period, therefore the primary diagnosis was considered to be sarcoidosis. The patient is still being followed-up without having any treatment.
Conclusions: Based on a rare manifestation, thyroid involvement in a patient diagnosed with sarcoidosis on the basis of clinical and pathological findings, we intended to underline in this report that sarcoidosis may involve any organ and this possibility should always be taken into account while monitoring the patients.

P21

Prevalence and risk factors for vitamin D deficiency in sarcoidosis

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Aim: Hypercalcemia, a common feature in sarcoidosis, is due to the excessive production of a Vitamin D metabolite, 1.25 $(OH)_2D$. Vitamin D efficiency in sarcoidosis though may not be appropriate. The purpose of this study was to assess Vitamin D levels in a sarcoidosis out-patient clinic compared to a mixed respiratory out-patient clinic population.
Material and methods: 64 sarcoidosis cases and 53 control cases with other than sarcoidosis respiratory diseases, matched for age and sex, were prospectively included in the study. Serum Vitamin D, 1.25 $(OH)_2D$, albumin, calcium, angiotensin converting enzyme were measured. Vitamin D was defined as deficient when < 20 ng/mL and insufficient when < 30 ng/ml. Clinical parameters for sarcoidosis cases were recorded.
Results: Overall 41/64 (64%) of sarcoidosis subjects had low Vitamin D levels. They were more likely to exhibit Vitamin D deficiency (39%) or normal Vitamin D levels (36%) than controls ($p = 0.018$). Vitamin D deficiency in sarcoidosis Group was associated with African-American race and radiological stage I disease. Regression analysis identified African-American race as the only significant factor predicting Vitamin D deficiency in sarcoidosis ($p = 0.034$). An inverse correlation between ACE and Vitamin D levels marginally insignificant was noticed ($p = 0.052$). 1.25 $(OH)_2D$ was significantly elevated in sarcoidosis cases compared to controls.
Conclusions: Vitamin D deficiency is prevalent in sarcoidosis. Severe Vitamin D deficiency is more prevalent in sarcoidosis patients than in patients suffering from other respiratory diseases. Among sarcoidosis patients, African-Americans are mostly in danger for hypovitaminosis D and should be monitored closely.

P22

Smoking related interstitial fibrosis — a new entity

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Introduction: Smoking related interstitial fibrosis (SRIF) was recently described as a distinct form of hyalinized interstitial fibrosis associated with emphysema and respiratory bronchiolitis. The diagnosis is often fortuitous and is not usually associated with significant respiratory symptoms. Functionally these patients may present obstruction and mild to moderate decrease DLCO. The most typical radiological findings in chest HRCT are well delimited subpleural emphysema, predominantly in middle and upper regions, associated with ground-glass opacities (GGO) and reticulation. The authors describe three cases of patients diagnosed with SRIF.
Case 1: 49 years old man, mechanic, smoker, asymptomatic, attended the ILD outpatient clinic due to radiological features (micronodules and GGO). For suspected DIP went through bronchoalveolar lavage (BAL) and transbronchial cryobiopsy (TBC), which revealed histological findings compatible with SRIF.
Case 2: 55 years old man, construction worker, smoker, asymptomatic, was sent to ILD clinic due to radiological features (emphysema, fibrosis and peripheral GGO). For suspected non-specific interstitial pneumonia, BAL and TBC were held, revealing findings consistent with SRIF.
Case 3: 60 years old man, tiler, smoker, with occasional exposure to birds, was referred due to blood-streaked sputum. The chest HRCT suggested features of RB-ILD and DIP pattern. BAL and TBC were performed, showing SRIF features.
Discussion: The authors wish to illustrate a distinct entity that needs to be recognized owing to a variable pathophysiology and prognosis, although the real natural history of SRIF is not full acknowledged.

P23

Efficacy of adalimumab in sarcoidosis patients who developed intolerance to infliximab

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Introduction: Tumor necrosis factor-alpha (TNF-α) inhibitors are regarded as the third-line therapy in sarcoidosis, the first choice generally being infliximab. To date, data regarding response to adalimumab in sarcoidosis patients intolerant to infliximab are lacking. The objective of this retrospective observational study was to establish if adalimumab could achieve stabilization or improvement of the disease in refractory sarcoidosis patients who developed intolerance to infliximab.
Material and methods: Sarcoidosis patients referred to St. Antonius Interstitial Lung Diseases Center of Excellence, Nieuwegein, The Netherlands, between January 2008 and April 2015 who switched from infliximab to adalimumab were included. Changes in organ function, inflammatory biomarker levels, and adverse events were retrieved from medical records.
Results: Out of 142 infliximab treated patients, 18 (13%) had to discontinue treatment due to antibody formation or severe adverse events and switched to adalimumab therapy. Organ function improved in 7 patients (39%), was stable in 6 patients (33%), and worsened in 5 patients (28%) after 12 months of treatment or after 6 months if evaluation after 12 months was not available (n = 4). In none of the patients biomarker levels of soluble interleukin-2 receptor (sIL-2R) deteriorated. Median decrease in sIL-2R was 3614 pg/mL. Most reported adverse event was infection (n = 10).
Conclusions: Adalimumab is an effective alternative for patients intolerant to infliximab. The switch to adalimumab achieved clinical improvement in 39% and stabilization in 33% of patients intolerant to infliximab. Further research is needed to develop guidelines on how to use adalimumab for sarcoidosis in terms of dosing regimen.

P24

The effect of MUC5b promoter polymorphism on bronchoalveolar lavage fluid characteristics in idiopathic interstitial pneumonias

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Introduction: A polymorphism in the *MUC5B* gene (rs35705950) is associated with susceptibility to fibrotic idiopathic interstitial pneumonias (IIP). MUC5B is a gel forming mucin and a major component of airway mucus.

Aberrant function or regulation of mucins is possibly an important component of mucosal infectious and inflammatory diseases. We investigated MUC5B protein levels in bronchoalveolar lavage fluid (BALf) in different forms of IIP and connective tissue disease associated interstitial pneumonia (CTD_IP). Furthermore, we assessed if *MUC5B* genotype influences BALf characteristics in IPF.
Material and methods: MUC5B levels were measured in 72 IPF, 24 FIP, 24 iNSIP, 20 CTD_IP patients and 52 controls. MUC5B was measured in BALf supernatant by ELISA. From IPF patients we collected smoking behaviour, age and lungfunction at diagnosis.
Results: MUC5B BALf levels were significantly higher in IPF, FIP, iNSIP and CTD_IP compared to controls. There were no significant differences seen between the diseases. MUC5B levels correlated with DLCO% predicted in IPF patients at diagnosis ($r = -0.45$; $p = 0.001$), the higher MUC5B, the lower DLCO% predicted. BALf MUC5B levels were significantly higher in ever smokers than in never smokers in IPF. MUC5B BALf levels were not dependent on the *MUC5B* rs35705950 genotype in controls or IPF. In IPF, *MUC5B* minor allele associated with significantly lower count of neutrophils ($p < 0.01$) and eosinophils ($p = 0.01$) in BALf. In controls no correlation between BAL cell profiles and the polymorphism was found.
Conclusions: IPF BAL cell profiles show that *MUC5B* minor allele carriers have lower inflammatory cell counts and might therefore be less prone to infections.

P25

Airway-centered interstitial fibrosis — rare interstitial lung disease: 2 case reports

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Airway-centered interstitial fibrosis (ACIF) is a new and rare interstitial lung disease (ILD) of unknown cause characterized by chronic cough and progressive dyspnea; history of inhaled exposure; propensity for women; occurrence in middle age; progressive peribronchiolar distribution of interstitial inflammation and fibrosis with bronchiolar metaplasia distinct from any form of ILD or idiopathic interstitial pneumonia (IIP). The majority of patients are non-responsive to corticotherapy with poor prognosis. We describe 2 female patients, 65 and 66 years old, presenting with chronic dry cough and progressive dyspnea. Both non-smokers, farmers and with a history of inhaled exposition to birds. Chest radiographs revealed diffuse reticulonodular infiltrates and bronchial walls thickening. ChestCT showed reticular fibrosis and ground glass infiltrates across the pulmonary fields in one and sparing the upper on the other. Bronchoalveolar lavage showed a small increase in lymphocytes/neutrophils in one, the ratio CD4/CD8 < 1 in both. The diagnosis was made by surgical biopsy revealing pericentrilobular lesions and linfoplasmocitary infiltrate compatible with ACIF. Both patients began systemic corticotherapy for 12 months. In one case the disease progressed with worsening symptoms, pulmonary functional tests and CT imaging even after starting combined imunosuppression with Azathioprine. The other patient improved with less symptoms and decreased area of disease on the CT image. These 2 cases presented similar clinical, radiological and pathological features as most of the few cases reported in literature. Our patients were non-smokers but were exposed to birds. The evolution varied with one improving after therapy and the other progressing even after combined imunosupression.

P26

Rare organ involvement in sarcoidosis: report of 10 cases

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Introduction/aim: Sarcoidosis is a multisytem disease with unknown etiology. Lungs are mostly affected. Lymph nodes, eye and skin involvements are seen according to the order of frequency. Other organ involvements are seen rarely. We aimed to take attention of diagnosis and treatment problems in sarcoidosis cases with rare organ involvement followed in sarcoidosis outpatient clinic.
Material and methods: 651 cases followed in sarcoidosis outpatient clinic were examinated retrospectively. Data of patients with sarcoidosis have rare organ involvement were gained.
Results: Eleven rare organ involvement was present in 10 patients (neurosarcoidosis (Guillain-Barre syndrome (GBS)), Bone marrow (BM), Thyroid (T), heart (H) (n = 2), gall bladder (GB), nodular splenic involvement (S) (n = 2), stomach/liver (S/L) (n: 2): musculoskeletal (M)). Nine cases were stage 1, one case was stage 2. The pulmonary diagnosis of cases were histologically except GBS (bronchial biopsy (n = 3); medistinoskop (n = 5)) wedge resection (n = 1)). The diagnosis of extrapulmonary organ involvement was by histopathological (BM, T, GB, M, S/L) and radiologically. hypercalcemia (n2), eye (n2), and skin (n = 2) involvement were obtained other than rare organs involvements. Treatment was began in four patients have life-threatening organ involvements (GBS, H, BM, S/L). Treatment has been completed in 2 patients with cardiac involvement. Patients still follw-up with no symptom.
Conclusions: Some Extrapulmonary involvement in sarcoidosis have a poor prognosis. Early diagnosis of EPS and prompt initiation of corticosteroid therapy with or without other immunosuppressants is crucial.

P27

Negative outcome of prednisone in possible idiopathic pulmonary fibrosis

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Introduction: The diagnostic classification "possible idiopathic pulmonary fibrosis (IPF) " is characterised by an inconsistent usual interstitial pneumonia (UIP) pattern on HRCT-scan and a UIP pattern in surgical lung biopsy. Therapeutic management in patients with "possible IPF" is challenging. The clinician must choose between either immunomodulatory agents or anti-fibrotic agents, but evidence is lacking.
Material and methods: A multi-centre cohort of 59 patients with "possible IPF" treated with prednisone were retrospectively analysed. Prednisone starting dose was 0.5 mg/kg/day and tapered to 0.15 mg/day/kg in six months. Patient demographics and serious adverse events (SAEs), defined as death and hospital admissions, were collected. Forced vital capacity (FVC) before start of therapy, baseline (start of therapy) and six months after start of therapy were evaluated.
Results: In 59 prednisone treated "possible IPF" patients, 22% had a SAE: twelve in the first three months on prednisone > 0. 3 mg/kg/day and two during the last three months on < 0. 3mg/kg/day prednisone. Patients had a mean decline of 8. 7% FVC before treatment and a further mean decline of 21% after treatment ($n = 32$; $p = 0.018$). The majority of patients were non-responders (69%) with FVC > 5% decrease or death within six months from baseline. Six former smoking patients with an additional histopathological desquamative interstitial pneumonia (DIP) component besides UIP, were responders with a mean increase of 6% FVC.
Conclusions: Patients with "possible IPF" demonstrated accelerated FVC decline and high incidence of SAEs during high dosed prednisone treatment. Presence of concomitant DIP histopathological pattern is associated with responsiveness to prednisone treatment.

P28

Microscopic polyangiitis: report of a clinical case

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Introduction: Vasculitis are a Group of diseases that cause necrosis and inflammation of the blood vessel wall which eventually culminates in it's destruction.
Case: 59 years male, chainsaw operator, ex-smoker, history of alcoholism, gout and anemia. Treated with Iron, pantoprazole and allopurinol. Observed in the ER by nausea, vomiting and mild peripheral edema for 15 days and epigastric pain with hemoptysis in the latter 24 hours. Physical examination: uremic breath, mucocutaneous pallor and mild edema to the ankles.

Afebrile, eupneic and normotensive. Cardiac auscultation and pulmonary unchanged. Analytically with anemia, acute kidney injury, PCR 1 and metabolic acidosis with partial respiratory failure. Urine II with proteinuria, erythrocyturia and glycosuria. Chest radiograph revealed bilateral alveolar pattern in the lower 2/3. Performed hemodialysis and empirically started methylprednisolone, cyclophosphamide and plasmapheresis. Next days without improvement in renal function but resolution of respiratory complaints. CT-Thorax showed interstitial densification and mediastinal lymphadenopathy and bilateral pleural effusion. Respiratory function test with DLCO 77. Kidney ultrasound revealed attenuation of the normal parenchymal-sinus differentiation in the left kidney. Renal biopsy showed crescentic pauci- immune necrotizing glomerulonephritis. Analytically with positivity for anti-MPO. Discharged medicated with cyclophosphamide 50 mg id, prednisolone 60 mg id at weaning. Outpatient observed in the 3rd month after discharge: no respiratory symptoms and CT-Thorax control without parenchymal changes. DLCO 71. Held switch to azathioprine 100 mg.
Conclusions: Systemic necrotizing vasculitides correspond to the majority of cases of diffuse alveolar hemorrhage of autoimmune cause. The serological study and renal biopsy are crucial for the differential diagnosis.

P29

A possible association between silicosis and sarcoidosis?

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Introduction: Sarcoidosis is a granulomatous inflammatory disease of unknown etiology, genetic, infectious and environmental factors have been proposed to induce. Silicosis has been suggested to increase rates of autoimmune diseases as well. In present study, we hereby report 8 cases of sarcoidosis who have silicosis secondary to organic dust inhalation.
Case: The study is conducted in a teaching hospital reference center for chest diseaes. Out of 616 sarcoidosis patients, who are being followed in our sarcoidosis outpatient clinic, 8 cases having inhalational dust exposure to silica crystalline were recorded retrospectively. All cases were male and the mean age was 40 (24–56). One case was asymptomatic whereas shortness of breath, cough and chest pain were the most common symptoms. Three patients were never smoker. Chest radiograms were staged as 3 in 5 cases. All cases had bilateral nodules, bilateral hilar and mediastinal lymphadenopathy in chest CT. One case had restriction in spirometry, ACE was high in 3 cases. Diagnosis was made by mediastinoscopy (n = 6), mediastinoscopy and lung biopsy (n = 1), gallium scintigraphy (n = 1). The median follow-up duration was 47 months. Radiological progression was recorded in one case whereas others remained stable.
Conclusions: Coincidence of silicosis and sarcoidosis may suggest that there could be a common pathogenesis. Occupational history is important in sarcodiosis and silicosis patients' may need assessments from this perspective as well.

P30

A case report with follicular bronchiolitis

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Introduction: Follicular bronchiolitis is a rare disease characterized by hyperplastic reactive lymphoid follicles and germinal center formation in the bronchioles wall histologically. It may be idiopathic or secondary to some causes such as connective tissue diseases, immunodeficiencies, malignancies, hypersensitivity or infection. We report a case with follicular bronchiolitis in order to discuss the diagnosis and treatment.
Case: A 57 year-old male patient was admitted to our clinic with sputum and fever continued for 6 months. He had been diagnosed as pnemonia and used antibiotics for 3 times in the last 6 months. He had smoked 50 pack/years, was fashion designer, was born and raised in Istanbul and had no other disease. Bilateral reticulonodular infiltrates significant in lower zones were detected on chest radiograph. On Chest CT, tree-in-bud signs and micronodules were detected. Sputum and bronchoscopic lavage samples did not yield mycobacteria or any other infectious agent. For definitive diagnosis, he underwent surgical biopsy and was diagnosed as follicular bronchiolitis. No etiologic factor was found in the detailed history. Clarithromycin and steroid treatment was started. In the first month, clarithromycin treatment was discontinued due to clinical improvement. In the fourth month, radiological right lung regressed significantly whereas a little progression was detected in the left lung. Clarithromycin was re-added to steroid therapy. ntinued. The patient is in 4 months of therapy is still in our follow- up.
Conclusions: Follicular bronchiolitis has been reported as case presentations in the literature. Treatment is planned due to underlying cause but there is no treatment protocol for idiopathic situations. Close monitoring of these patients is important.

P31

Organizing pneumonia and pulmonary fibrosis secondary to Sjögren Syndrome: case report

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Introduction: Sjögren syndrome is a systemic chronic inflammatory disorder associated with lymphocytic infiltration of exocrine glands. In addition, multiple extranglandular features may develop, including pulmonary involvement.
Case: 62 years old female, unemployed, non-smoker, with history of right mastectomy, hypothyroidism, depression and osteoporosis. Treated with alprazolam, venlafaxine, levothyroxine, alendronic acid, colecalciferol. Patient referenced with dyspnea complaints to moderate exertion, wheezing and asthenia in 2010, also xerophtalmia. Physical examination showed mild hypertrophy of right parotid. Thoracic radiography with lower bilateral reticular pattern and HRCT with interstitial pattern and ground glass opacities more evident in the middle lobe and lower lobes. Analyzes with positive ANA 1/1280 granular pattern, FR and anti-SSA. Respiratory functional study with restrictive pattern. No changes in blood gases. Transthoracic biopsy performed which revealed organizing pneumonia but the patient did not want to start treatment at this time. Sent also to a Rheumatologist who made diagnosis of primary sjögren syndrome. At 6 months follow-up, the patient maintained respiratory complaints and accepted start treatment with prednisolone 20 mg for 1 year, with improvement of respiratory complaints. Reevaluation HRCT at 2012 showed progression to pulmonary fibrosis (craniocaudal gradient and subpleural space achievement) with diffuse ground glass. In 2015, further deterioration of respiratory complaints with hypoxemia and HRCT showed worsening fibrosis. Started prednisolone 50 mg, azathioprine 50 mg id and LTOT, maintaining follow-up at this time.
Conclusions: Sjögren's syndrome is a common auto-immune disease. The respiratory system is frequently involved with 10% of the patients developing a clinically significant lung disease.

P32

Vertebral fracture — initial presentation of sarcoidosis

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Introduction: Sarcoidosis is an inflammatory disorder characterised by granuloma infiltration of two or more organs. Exclusion of other aetiologies, mainly infectious, autoimmune and neoplastic is mandatory in a demanding process as demonstrated in this case report.
Material and methods: Review of clinical file and scientific literature.
Case: A 59 year old healthy non-smoker female was admitted for investigation of an L2 fracture with lytic lesions after a low energy trauma. No previous infections, environmental exposure or travel history were determined. Apart from lower back non-irradiating pain, physical examination was unremarkable. A tomography revealed multiple organ infiltration: lungs, liver, lytic bone lesions (vertebrae, sternum, iliac and ribs) associated with cervical, mediastinal and abdominal lymph nodes. Biopsies of cervical lymphadenopathies and iliac bone showed chronic inflammatory tissue with granulomas. Neoplastic aetiology was excluded after colonoscopy, endoscopy, mammogram, thyroid ultrasound and complete gynaecologic and dermatologic studies. Interferon Gamma Release Assay, blood cultures, infectious and autoimmune serologies excluded other causes of systemic granulomatous disease. Bronchoalveolar lavage cultures were negative and

CD4/CD8 ratio was technically undeterminable. Treatment with corticosteroids was started pending radiologic revaluation.
Conclusions: The authors present a case of widespread sarcoidosis with pulmonary, lymph nodes, liver and bone involvement. Bone involvement can be seen in < 10% of sarcoidosis patients, most commonly in the form of cystic osteitis. Vertebral lytic lesions are noted in < 1% of extrapulmonary sarcoidosis making this a rare manifestation of sarcoidosis. Long term follow up and positive response to corticosteroids will be determinant to absolutely exclude alternative diagnosis.

P33

Organizing pneumonia as an isolated manifestation of anti-synthetase syndrome

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Introduction: The usual clinical manifestations of anti-synthetase syndrome are myositis, fever, polyarthritis, raynaud phenomena and interstitial lung disease. Immunologically it presents with positive anti-tRNA synthetase antibodys. This case report shows an unusal manifestation of this syndrome.
Material and methods: We made a summary of the case report, including diagnostic exams.
Results/case report: 58-year-old woman, with a clinical background oh high blood pressure and cerebral ischemic event. ANA autoantibodies always positive in previous analysis, without autoimmune disease criteria. Admitted for sense of fatigue, effort dyspnea, dry cough and fever. Pulmonary auscultation showed bibasal crepitations. Chest radiography: bibasal condensations; blood analysis: neutrophilic leukocytosis, PCR 6. 82 mg/dL. Clinically improved after antibiotherapy. Readmitted 2 weeks later for the same symptoms, hypoxemic (pO2 59 mm Hg), when she performed thoracic CT-scan: bibasal consolidations with air bronchogram; pulmonary-function tests: small-airways obstrution; positive anti-EJ antibody; bronchofibroscopy with normal macroscopic aspect, bronchoalveolar lavage with 700cells/mm3, 70% linfocites (CD4/CD8 = 0.22), 24% macrophages, 6% eosinophiles, 0% neuthrophiles, BK search was negative; Normal eletromyography. The patient started corticotherapy (prednisolone 1mg/Kg/day) with rapid clinical improvement. Thoracic CT-scan after 3 month on corticotherapy showed only some pulmonary fibrosis.
Conclusions: Though anti-synthetase syndrome is a systemic disease, it may present with isolated pulmonary involvement, which is the main prognostic determinant in this patients. Autoantibodies may represent a precious help on the diagnostic aproach of patients with isolated interstitial lung disease.

P34

Non-specific interstitial pneumonia — clinical associations and evolution

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Introduction: Non-specific interstitial pneumonia (NSIP) is the second most common pattern of idiopathic interstitial pneumonias. Although natural history of NSIP is not yet fully established and its evolution depends on the underlying etiology, NSIP usually comprises a good prognosis.
Aims: Characterization of patients with NSIP.
Material and methods: Retrospective analysis of patients with NSIP, encompassing etiology, clinical, functional and imagiologic evaluation, as well as evolution characterization.
Results: Included 57 patients, with a mean (± SD) age of 60 (± 12) years and 78. 9% female. The majority (54. 4%) was non-smoking. Possible etiologies for NSIP pattern were related in most patients (61. 4%, n = 35) with CTD and in the remaining with drug toxicity (n = 10), chronic HP (n = 10) and 2 cases were considered idiopathic. Regarding CTD patients, 13 had systemic sclerosis (SSc), 10 rheumatoid arthritis, 4 dermatomyositis/polymyositis, 3 Sjögren's syndrome, 3 mixed connectivite tissue disease and 2 systemic lupus erythematosus. Drugs associated with NSIP were statins (n = 7) and metformin (n = 3). At baseline functional assessment: mean (± SD) FVC of 97 (± 22.6)%, FEV_1 of 97.3 (± 20.1)% and TLC of 93.2 (± 184)%. The majority of patients (n = 41) had decreased DLCO (65.3 (± 20.2)%). Between CTD patients, there were lower values of FVC in SSc ($p < 0.05$). In 47.4% of NSIP cases, immunosuppressive therapy was introduced. Only 5 patients (10.2%) died during the follow-up (in 2cases cause of death was not related to NSIP).
Conclusions: In this series of cases with NSIP, it was found that the CTD were the main responsible etiology, and few cases were idiopathic. Major functional impairing was reflected in a decrease in DLCO.

P35

Low apoptosis rate of alveolar lymphocytes (AL) in sarcoidosis is characteristic only for active forms of the disease

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Aim: Examination of AL apoptosis frequency in sarcoidosis, with relation to the clinical presentation, i. e. in patients subdivided into Loefgren's Syndrome (LS), chronic stable sarcoidosis (SC) and chronic progressive sarcoidosis (PS).
Material and methods: BAL of 103 sarcoidosis patients and 17 controls was examined for: a) AL apoptosis in flow cytometry (sub-G1 peak of cell cycle); b) TUNEL assay; c) AL staining for BCL2 family, caspase-3, and death receptors/ligands (Fas, DR3, DR4, TNFR1, FasL, TRAIL); d) cytoimmunology. Additionally, in 7 PS patients AL apoptosis was revaluated after 3-6 months of systemic corticosteroid treatment.
Results: AL apoptosis rate was lower ($p < 0.05$) in LS and PS, but not in CS, than in controls (e. g. sub-G1 peak for LS: 0.3 ± 0. 2%, CS: 0.8 ± 0.3%, PS: 0.4 ± 0.4%, controls: 0.9 ± 0. 6%, median ± SEM); parallel TUNEL data were obtained. AL in sarcoidosis was characterized by lower expression of TNFR1 (significant for LS and PS), FasL and TRAIL (significant for all), caspase-3 (significant for LS). Steroid treatment in PS patients resulted in remarkably increased percentage of apoptotic AL (incl. cases of "mitotic catastrophe" in cell cycle) and decline of BCL2+ cells. Moreover, AL apoptosis rate in sarcoidosis was negatively correlated with BAL lymphocytosis ($Rs = -0.49$, $p < 0.01$) and AL activation (percentage of CD3+HLA- DR+ cells: $Rs = -0.28$, $p < 0.002$).
Conclusions: AL apoptosis rate in sarcoidosis is declined only in active forms of the disease. The possible mechanisms include high expression of BCL2 and low of death receptors and their ligands. AL infrequent apoptosis is responsible for development of lymphocytic alveolitis in the disease. Corticosteroid use benefit in chronic progressive sarcoidosis induces high rate of AL apoptosis.

P36

Do alveolar lymphocytes (AL) present antigens? Evidence from BAL cell immunotyping in interstitial lung diseases (ILD)

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Introduction: Antigen presenting cells (APC) participate in ILD by priming specific T cells, regulating inflammatory response and disease outcome. Local APC system includes dendritic, Langergans and B cells, as well as alveolar macrophages. Interestingly, we have previously shown that AL produce APC-linked cytokine, IL-27. There is a question, if T cells possess a local ability to present antigens.
Aim: Immunotyping of APC markers on lymphocytes (AL) harvested by BAL from ILD patients.
Material and methods: AL originating from BAL carried out in sarcoidosis, PS, idiopathic pulmonary fibrosis, IPF, nonspecific interstitial pneumonia, NSIP, exogenous allergic alveolitis (EAA) and controls (n = 33, 17, 10, 9, 7) were immunotyped for HLA-DR, CD80, CD86 (costimulatory B7 molecules) CD83 and CD1a with use of flow cytometry.

Results: AL, Th and Tc cells, in similar extension, express mostly HLA-DR, CD80 and/or CD86. The HLA-DR+CD80+CD86+ coexpression occurs in AL of all tested Group s and varies from 0 to 16% of CD3+ cells. It is increased in IPF, as compared to controls, while PS and EAA is characterized by significantly lower percentage of CD80+CD86+ cells (e.g. CD4+HLA -DR+CD80+CD86+: 2.6 ± 1. 4% in PS and 1.7 ± 1.1% in EAA as compared to 8.3 ± 3.7% in controls; median SEM, $p < 0.05$). AL CD80+CD86+ percentage was negatively correlated with BAL lymphocytosis and number of AL with T effector phenotype (CD3+CD27-CD28-); there was no relation to patients clinical data, including lung function test results.
Conclusions: 1. AL phenotype suggests T cells to be active in ILD as APC. 2. Frequency of AL APC-linked phenotype is declined in ILD patients with high BAL lymphocytosis.

P37

Lipidomics in sarcoidosis — preliminary report

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Introduction: The aim of this study was to determine the use of the lipid profile of patients with sarcoidosis and compare it with healthy subjects. We assume that lipid profile of serum in sarcoidosis differs from the control subjects lipid profile.
Material and methods: Serum was collected from 9 patients with II stage of sarcoidosis and 5 control subjects. In our studies proton nuclear magnetic resonance (NMR) spectroscopy of lipid extracts was used. NMR spectra were collected using 400 MHz spectrometer and standard one pulse sequence. Lipids were extracted from serum before analysis using modified Blight and Dyer method and dissolved in deuterated chloroform. Thirty four NMR signals of lipid compounds were analyzed. Partial least square discriminat analysis (PLS-DA) with Pareto scaling were used to analyzed lipid profile.
Results: Univariate t-test analysis show significant differences in NMR signals of both esterified and free cholesterol and fatty acids ($p < 0.05$). For analyzing lipid profile discriminant analysis was applied. Obtained PLS-DA model consisted of three components and very good explain the data and also predict the data. Discriminant analysis correctly classified patients according to their Group s for 100% of sarcoidose and 100% of control. Lipidomics indicated significant differences in phosphatidylcholine, triglycerides, fatty acids and sphigomyelin two unassigned compounds (4. 94ppm, 514ppm) levels.
Conclusions: Lipidomics is able to differentiate sarcoidosis patients and control subjects based on lipids profile of serum. Our investigation demonstrated that this technique can offer also insight into the lipid states of sarcoidosis.

P38

CVID pretending sarcoidosis as a result of primary immune deficiency with pulmonary predominance in adulthood: a case study

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The case report presents 33-year-old female patient who had chronic bilateral lymphadenopathy with pulmonary involvement *sub forma* small, scattered granuloma and patchy area of peripheral fibrosis, visible in chest CT. Before 2013 the patient was misdiagnosed as sarcoidosis, luckily without treatment due to lack of recommendation (mildly visible changes in PFT, with no extrapulmonary manifestations).
Firstly, the patient was suspected of persistent pulmonary sarcoidosis, without spontaneous remission.
Due to worsening in pulmonary function test, which showed mild restriction with lowered DLCO the patient was admitted to our centre — with ILD focused unit. After comprehensive examination, it turned out that despite lung disease, the only malady is BMI below 17, than she was tested to Ig M, G, A levels — which were extremely lowered. To reassure our team's suspicious, we perform VATS. Histopathological sample was ultimately classified as not typical sarcoid granuloma, but poorly formed granuloma like in inflammatory response linked to immune deficiency syndrome. Having achieved those results, and excluding others causes we started treatment with immunoglobulin supplementation — with total regression of the disease in the thorax.
CVID as a primary immune deficiency (PID) syndromeis associated with usually early respiratory tract involvement, and is diagnosed in childhood rather, due to recurrent respiratory infections or gastric manifestation. But being aware of PID delayed manifestations, as well as sarcoidosis linked to IgA deficiency syndrome, nowadayswe should put an extreme attention to perform proper diagnosis of pulmonary changes including rare disorders.

P39

Detection of anti DFS70 antibodies in patients with interstitial lung disease (ILD) with and without connective tissue disease (CTD)

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Introduction: Anti-DFS70 antibodies, corresponding to the dense fine speckled ANA indirect immunofluorescence pattern in HEp-2 substrates, have been observed in chronic inflammatory conditions, cancers and even in healthy individuals but only in a small percentage of patients with systemic autoimmune rheumatic diseases (SARD). Aim of this study: To investigate the frequency of serum anti- DFS70 in patients with ILD and a possible correlation with ANA status and the presence of CTD.
Material and methods: 113 patients with ILD (70 IPF and 43 NSIP), 50 healthy controls and 36 scleroderma patients as negative control were studied. 14 patients had or developed CTD during 24 months of follow up (1 IPF and 13 NSIP). Serum anti-DFS70 at baseline was measured by ELISA (Medical and Biological Laboratories Co., Ltd., Japan), and the cut-off for positivity was set at 400 U/mL.
Results: ANA were positive in 20/43 (47%) NSIP patients and 22/70 (31%) IPF patients. All healthy controls were ANA negative and 36% anti-DFS70 positive. All scleroderma ILD patients were ANA positive and only 6% anti-DFS70 positive. Anti-DFS70 were positive in 7/43 (16%) NSIP patients and 15/70 (21%) IPF patients. Among ANA (+) ILD patients (42), only 6 (14%) were anti-DFS70 positive and among ANA (−) ILD patients (71), 16 (23%) were anti-DFS70 positive. 8/36 (22%) of ILD patients with ANA (+) and anti-DFS70 (−) and 1/6 (16%) of ILD patients with ANA (+) and anti-DFS70 (+) had or developed CTD over time, all of them had NSIP.
Conclusions: Anti-DFS70 positivity is more frequent in ANA negative healthy subjects. ANA positivity combined with anti- DFS70 negativity seems to be associated with CTD in NSIP patients.

P40

Diagnostic usefulness of [18f]FDG-PET/MRI in patients with sarcoidosis: pilot study

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Introduction: PET/MRI is a new hybrid imaging technic which enables - simultaneous whole body molecular magnetic resonance and metabolic positron emission tomography made in one study.
Aim: The purpose of this study is the evaluation of the impact of [^{18}F] fluorodeoxy-d-glucose (18F-FDG) positron emission tomography (PET) /3T whole body magnetic resonance (MR) hybrid use on diagnosis and staging of sarcoidosis patients.
Material and methods: FDG-PET/MR was performed in 26 patients (F/M: 10/16, mean age: 45 (25−69), with computed tomography (CT) diagnosed and histologically confirmed pulmonary sarcoidosis. PET scans were obtained 60 min after injection of 295 ± 45 MBq [^{18}F]FDG). All patients underwent ^{18}F-FDG PET/MR and CE-MRI (Contrast Enhanced Magnetic Resonance) of the thorax at 3T in the prone position with proper MR sequences. Quantitative assessment was performed by calculation of SUV_{MAX}. Disease staging was performed independently by two nuclear medicine and one radiology specialist.
Results: PET/MRI confirmed metabolically active disease in the lesions localized by CT in all patients. Fifteen (57%) patients were classified as

phase I, and 11 (43%) as phase II pulmonary sarcoidosis. Increased 18F-FDG uptake in PET/MR study, with SUV_{MAX}: 2.43–9.00, as a new additional metabolically active foci were found in: abdominal lymph nodes in 8 (30%) patients, cardiac left ventricule — in 1 (3%), musculo-sceletal system in 1 (3%) patient. Additionally, in seven patients with nonspecific neurological symptoms slight CNS changes were found on MR images.
Conclusion: FDG-PET/MRI might be a promising tool in early and comprehensive diagnosis of sarcoidosis relevantly influencing decision-making process.

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Risk factors for increased mortality in sarcoidosis

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Introduction: The mortality rate of the sarcoidosis is 5–10% however little are known for the parameters that estimate this rate. Aim to determine risk factors that could increase mortality of the disease.
Material and methods: 122 patients (43 men, mean age 48.65 ± 13.3 years) with biopsy proven sarcoidosis were studied retrospectively. Lung function tests including total lung capacity (TLC) and diffusion capacity (DLCO), surface electrocardiogram, cardiac ultrasound, 24-hour Holter monitoring, BNP and SACE were examined. Mean heart rate, maximum and minimum heart rate and time domain indices of heart rate variability (Standard Deviation of RR-SDRR, RMSDD, pNN50) were calculated from the 24 hour Holter while evidence of ventricular arrhythmias were noted. Cardiac sarcoidosis was detected if there were abnormal finding on cardiac ultrasound, ECG and/or holter and cardiac MRI.
Results: During a median 58.89 ± 15.75 month follow-up, there were 10 deaths (8.2%). 5 patients died due to cardiac and 5 due to other causes. Cardiac involvement was estimated in 40 patients (32.8%). TLC < 80% of the predicted was presented in 29 (23.77%) while isolated DLCO reduction was presented in 21 (17.21%) patients. In the multivariate analysis, standard deviation of all normal-to-normal RR intervals < or = 90 ms, TLC < 80%, DLCO < 80% of the predicted and the presence of cardiac involvement were found to be independent risk predictors of all causes mortality.
Conclusions: Increased risk of mortality and cardiac death could be predicted in patients with sarcoidosis by evaluating the cardio-respiratory function. TLC and/or DLCO reduction, the presence of cardiac involvement and the standard deviation of RR intervals were independent predictors of all causes mortality.

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Polymorphism of activating *FCGR* genes in an etiopathogenesis of sarcoidosis

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Introduction: We have previously presented evidence that the polymorphism of the *FCGR3A* gene, encoding the receptor for Fc fragment of immunoglobulin G IIIa (Fc*γ*RIIIa) plays a role in the enhancement of circulating immune complexes (CIs) with the occurrence of mycobacterial heat shock proteins in patients with sarcoidosis (SA). The immunocomplexemia might be caused by decreased affinity of CIs to Fc*γ* receptors, with the subsequently decreased receptor clearance by immune cells. In the present study we examined whether the polymorphisms of other related genes (*FCGR2A, FCGR2C, FCGR3B*) encoding other activatory Fc*γ* receptors, could have a similar effect.
Material and methods: Thus, we analysed polymorphism of *FCGR2A, FCGR2C*, and *FCGR3B* in 104 SA patients and 110 healthy volunteers using PCR-SSP.
Results: Our study found significantly lower percentage of *FCGR2A* and *FCGR2C* with a concomitant increase of less functional variants of these genes in Stages I/II, than in Stages III/IV of SA. There was no aberration in *FCGR3B* allele/genotype frequencies.
Conclusions: We conclude that the *FCGR2A* and *FCGR2C* polymorphisms may also contribute to immunocomplexemia present in SA. The assessment of *FCGR* genes could become a tool in presaging a clinical course of sarcoidosis and in its personalized therapy.

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Bronchial and peritoneal sarcoidosis in a 30 y/o asymptomatic woman: case report

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Sarcoidosis (SA) is a multisystemic, granulomatous disease with unknown etiology. Infectious and non-infectious, genetic factors, as well autoimmunity have been considered as potential etiologic causes. Pulmonary involvement is the most prominent, however other locations are not infrequent. We have reported a case of a 30 years old Caucasian female, diagnosed with peritoneal sarcoidosis. The patient was asymptomatic, and was undergoing diagnostic procedures due to infertility. Transvaginal ultrasonography revealed non- characteristic changes localized proximal to the ovary and presence of a little amount of free fluid inside the peritoneal cavity. Due to subsequent high levels of serum CA-125 marker a suspicion of asymptomatic ovarian cancer was made. Results of histological examination of samples obtained from a diagnostic laparoscopy showed non-caseating granulomas in the peritoneum of the abdominal cavity, ovaries, oviducts, uterine body, and large intestine. Tuberculosis and carcinoma were excluded as a potential etiology of these findings. HR-CT scan of lungs showed no pathology but a diagnostic fiberoscopy with biopsy of the wall of the bronchi showed presence of sarcoid granulomas. This case has a potential scientific as well as clinical value, as it describes a case of peritoneal sarcoidosis, which is an extremely rare extrapulmonary manifestation of the disease, without any parenchyma lung involvement. The elevated level of CA-125 in sarcoidosis is a subject, which deserves a further study on a larger number of patients, as it has already been reported in a few cases.

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Influence of contrast enhancement CT and experience in chest imagining on the assessment of chest lymph nodes in sarcoidosis patients

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Aim: The aim of the study was retrospective analysis of chest CT evaluations of 40 patients with sarcoidosis.
Material and methods: Examinations consisted of non-enhanced (non-CE) and contrast-enhanced (CE) acquisitions that were performed at the same time at each patient, using the same CT unit, CT protocol with the same team of radiologists (less and more experienced in chest imagining). Significance of differences in parametric data (lymph nodes diameters, levels) were tested using ANOVA. Concordance between observers regarding of parametric data was determined using ICC, non-parametric data was assessed using Cohen's .
Results: A total analysis included 40 non-CE and 40 CE datasets for ANOVA analysis, 80 non-CE and CE datasets for comparisons between radiologists and residents. There were no significant differences between „ chest"radiologists and residents, apart from the dimension of the 4R node, but the number of affected node levels was found significantly higher when evaluated on CE images, within significant difference between all observers.
Conclusions: The application of contrast medium has a limited influence on the quality of general assessment of the chest lymph nodes in sarcoidosis patients, regardless experience of the observer. The necessity of contrast enhancement for chest lymph nodes imagining is still debated due to its nephrotoxicity. With corresponding clinical data of sarcoidosis common recommendation for using contrast enhanced CT for evaluation chest lymph nodes seems to be weak. CT imagining is usually important to find the best place for EBUS provided biopsy which is visible good enough in non CE CT.

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Age, forced vital capacity and anxiety sensitivity as predictors of dyspnea intensity in sarcoidosis

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Introduction: Sarcoidosis is a chronic multisystem disease of unknown etiology. In spite of the high prevalence of depression and anxiety in sarcoidosis, relatively little is established regarding the potential impact of psychological variables on respiratory symptoms reported in this disorder. Although every organ can be involved, the lungs are affected most often and dyspnea is one of the most frequents complaints in patients with sarcoidosis. There is some evidence that anxiety and anxiety sensitivity exacerbate dyspnea associated with pulmonary disease, but little is known about psychological factors contributing to dyspnea in sarcoidosis. Therefore, the aim of the present study was to investigate factors predicting dyspnea severity in sarcoidosis.
Material and methods: 110 patients (mean age 45. 3 years) were enrolled. They underwent spirometry and the following questionnaires: Hospital Anxiety and Depression Scale and Anxiety Sensitivity Index-3, MRC and items assessing demographic variables and symptoms.
Results: Dyspnea intensity was found to be related with higher age, physical concerns subscale of ASI (ASI_ph) and with lower FVC. Linear regression showed that FVC predicts the dyspnea intensity as long as the model lacks ASI_ph variable. The whole model explained about 18% of MRC dyspnea intensity variance, with age and ASI_ph as significant predictors explaining 7. 51% and 6. 50% of dyspnea variance respectively and FVC as non-significant predictor.
Conclusions: The finding that fear of physical sensations (ASI_ph) predicted more severe subjective dyspnea than FVC call for developing strategies to measure and reduce high levels of anxiety sensitivity to improve functioning in dyspneic patients with sarcoidosis..

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The role of PET in lung sarcoidosis: preliminary results from Greece

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Introduction: In sarcoidosis the ability to unveil unsuspected and persisting inflammatory activity (granulomas) may add in patient evaluation. 18F-fluoro-2-deoxyglucose (18FDG) positron emission tomography (PET) chest computed -tomography (CT) has been reported useful in the estimation of sarcoidosis activity.
Aim: To investigate the role of 18FDG/PET/CT in the evaluation of intrathoracic active disease and its relationship to functional and clinical findings in pulmonary-sarcoidosis-patients.
Material and methods: Sarcoidosis patients referred to our department between 2012 and 2015 were prospectively investigated by chest x-ray, CT, pulmonary function testing and 18FDG/PET/CT in addition to systemic investigation. Results One hundred twenty four consecutive sarcoidosis patients, 49.1% treatment-naïve, a median age of 52 years, Scadding radiographic stage 0, I, II, III, IV of 15.2/43.8/33/5.4/2.7, median FVC, Tiffeneau, DLCO, TLC predicted of 95.6%, 79.12%, 81%, 89.5% were studied. Female were 55.3% and no/ex-smokers 89.8%. Median diagnosis duration (IQR) was 2 years (0−8) and median follow-up (IQR) 13.5 (8−19) months. Acute and chronic disease was 51. 61 and 29.83% respectively. Based on 18FDG/PET/CT, 15.9% had thoracic disease only, 5.6% extrathoracic disease only and 68.2% had both thoracic and extrathoracic disease. Concerning intrathoracic 18FDG/PET/CT findings, bilateral hilar, subcarinal, subaortic lymph nodes and bilateral lung parenchyma were the most common sites of disease activity (61.3%, 52.4%, 32.35%, and 33.06% respectively). Pulmonary parenchyma active disease patients expressed lower FEV_1, FVC, DLCO and TLC ($p < 0.001$, $p = 0.001$ $p = 0.002$ and $p = 0.17$) and presented more often cough ($p = 0.045$).
Conclusions: 18FDG/PET/CT lung active disease was observed in 85% of patients and was associated with functional and clinical parameters.

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Myelodysplastic syndrome in idiopathic pulmonary fibrosis: dangerous liaisons

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Introduction: Comorbidities have an unfavorable effect on outcome of both myelodysplastic syndromes (MDS), and idiopathic pulmonary fibrosis (IPF). IPF is an irreversibly fibrotic disease and acute respiratory distress syndrome (ARDS) dramatically complicates its course. The constellation of both has been encountered in short-telomere syndromes with TERT or TERC germline mutations.
Material and methods/aim: To examine clinical characteristics, genetic background, and outcome of a small cohort of pulmonary fibrosis-MDS patients. PF-MDS patients diagnosed in 2015 underwent genetic testing for TERT and TERC sequencing after written informed consent. Demographic, clinical, laboratory, genetic characteristics and survival were reported.
Results: Five PF-MDS patients, 4 male, median age of 80-years, 60% ex-smokers, FVC = 85. 65, TLC = 64.25, DLCO = 62.1% predicted, Ht% = 33.9%, MCV = 105.6, WBC = 4250, PLTs = 171,000 were studied. IPF diagnosis predated MDS in 3. One patient presented refractory anemia (RA) and one RA with ring sideroblasts with low International prognosis scoring system (IPSS) score whereas 3 presented RA with excess blasts with intermediate-1, intermediate-2 and high IPSS scores respectively. TERT or TERC germline mutations were not identified. Two of 3 azacitidine-treated patients developed leukemia. All treated plus one untreated-patient (80%) succumbed to fatal ARDS upon IPF event during hospitalization for febrile infectious episode, 13 (9.75−14) months post-MDS-diagnosis and 9 (1−9) months post-MDS-treatment initiation.
Conclusions: MDS and IPF may co-exist without TERT or TERC germline mutations in aged patients. Outcome may be precipitated by fatal ARDS upon IPF triggered by infections in patients with high IPSS-score necessitating frequent hospitalizations and receiving aggressive myelosuppressive treatment.

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Cyclooxygenase-2 and miR-16 expression in pulmonary sarcoidosis

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Introduction: Elevated COX-2 activity is associated with the development of chronic lung diseases, including sarcoidosis. It was confirmed, that *COX-*

2 is targeted by the miR-16 leading to downregulation of *COX-2* expression by altering mRNA stability. This mechanism is claimed as a key posttranscriptional COX-2 regulation.

Aim: The aim of the study was to examine expression pattern of *COX-2* mRNA in relation to miR-16 expression in sarcoidosis patients.

Material and methods: Expression evaluation of both: *COX-2* and miR-16 was performed by qPCR method in bronchoalveolar lavage (BAL) cells and peripheral blood (PB) lymphocytes in sarcoidosis patients (n = 61) and control Group (n = 30). Analysis of *COX-2* mRNA revealed down-regulation (RQ < 1) in BAL fluid and in PB lymphocytes in patients and in control Group s with insignificantly lower *COX-2* expression in patients in BAL fluid ($p > 0.05$ Mann-Whitney U-test) and significantly higher *COX-2*expression in patients in PB lymphocytes ($p = 0.003$ Mann-Whitney U-test).

Results: Analysis of miR-16 expression in BAL fluid and PB lymphocytes revealed its upregulation (RQ > 1) in both biological material with insignificantly higher expression of miR-16 in BAL fluid and PB lymphocytes in patients ($p > 0.05$ Mann-Whitney U-test).

In total Group of sarcoidosis patients in BAL fluid we observed a trend towards negative correlation between *COX-2* and miR-16 expression in patients with II-IV radiological stage ($p > 0.05$, $r = -0.133$ Spearman's rang correlation test) and in patients with chronic form of disease ($p > 0.05$ $R = -0.02$ Spearman's rang correlation test) with reduced expression of *COX-2* in relation to miR-16.

Conclusions: Results suggest that reduced expression of mRNA *COX-2* in patients BAL fluid is associated with elevated expression of miR-16. Expression of *COX-2* mRNA in patients with pulmonary sarcoidosis may be directly controlled by miR-16.

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