Clinical perspectives in several interstitial diseases

P3740
Late-breaking abstract: Pathological analysis of acute exacerbation of idiopathic pulmonary fibrosis (IPF)
Yuh Fukuda1, Mika Terasaki2, Mikako Takahashi1, Shinobu Kunugi1, Yasuhiro Terasaki1, Hirokazu Urushiyama1, Arata Azuma2.
1Department of Analytic Human Pathology, Nippon Medical School, Bunkyo-ku, Tokyo, Japan; 2Department of Pulmonary, Infection and Oncology, Nippon Medical School, Bunkyo-ku, Tokyo, Japan

The patients with IPF show occasionally acute exacerbation in their clinical courses. However the details of pathology of acute exacerbation of IPF are not well investigated.

Patients and methods: We studied clinico-pathologically 15 autopsy cases of acute exacerbation of IPF. The clinical symptoms of acute exacerbation were acute respiratory distress syndrome, but multiple organ failure (MOF) was not associated. The intervals of acute exacerbation and death were 3 days to 6 months. The triggers of acute exacerbation were the infection in 3 patients, myocardial infarction in 3 patients, surgical operation in 4 patients, chemotherapy for cancer in 1 patient and unknown in 7 patients. All patients were treated with steroid. Autopsied lung tissues were observed with light microscopy and immunohistochemistry.
P3741 Interstitial lung disease associated by gemcitabine chemotherapy in non-small lung cancer patients: Analysis based on the data in practical use with confirmed denominator
Yukiko Miura, Yoshimobu Sato, Yuji Minegishi, Arata Azuma, Akihiko Gemma.
Nippon Medical School, Tokyo, Japan

Background: Gemcitabine (GEM), which is often used for non-small cell lung cancer (NSCLC), was reported to induce interstitial lung disease (ILD) in Japan. Presently, all-case postmarketing surveillance came to be generally conducted after the launch of new anti-cancer drug to investigate less-frequently but serious adverse drug reactions such as ILD in Japan. However, all-case surveillance had not been conducted for GEM and there is few data of GEM-induced ILD incidence in NSCLC practical use with confirmed denominator.

Methods: To investigate incidence, severity, and risk factors of ILD in NSCLC patients with GEM chemotherapy in single institute.

Results: We conducted a retrospective observational study of all NSCLC patients who received GEM in our hospital through medical records. Recorded chemotherapy for interstitial lung disease (ILD) in NSCLC patients collected from 1997-2010.

Conclusions: Incidence rate of GEM-associated ILD in NSCLC patients was 2.9% in this study. Most of the patients who developed ILD had a good prognosis but one of them had a fatal outcome. We weigh the difference between our result and previous reports of GEM-associated ILD.

P3742 Lymphoid hyperplasia and eosinophilic pneumonia as histologic manifestations of amiodarone-induced lung toxicity
Brandon T. Larsen1, Laszlo T. Vaszar2, Henry D. Tazelaar3, Thomas V. Colby3.
1 Pathology, University of Arizona, Tucson, AZ, 2Medicine, 3Laboratory Medicine and Pathology, Mayo Clinic Arizona, Scottsdale, AZ, United States

Background: Amiodarone use is often limited by lung toxicity. Amiodarone lung disease (ALD) classically manifests as organizing pneumonia with intra-alveolar foamy macrophages, but other patterns may occur. Herein we report two previously unreported patterns of ALD, lymphoid hyperplasia (LH) and eosinophilic pneumonia (EP).

Methods: We identified patients with LH or EP as a prominent feature among 75 cases of probable ALD from the authors’ teaching files collected from 1997-2010. Clinical history and wedge biopsies were reviewed.

Results: Twelve patients (7 men) met inclusion criteria. Median age was 71 years. Amiodarone dose was known in 10 cases (median 200mg/d). Treatment duration was known in 8 cases and ranged from 1 to 9 years. Thoracic imaging showed diffuse infiltrates concerning for ALD. Histologic review revealed intra-alveolar foamy macrophages in all cases. Eight cases prominently displayed patterns of LH including diffuse LH (7), follicular bronchiolitis (5), lymphocytic interstitial pneumonia (2), and lymphocytic perivascular cutting (2). Two showed features of acute EP including diffuse alveolar damage with abundant eosinophils. Two showed features of chronic EP including interstitial pneumonia with abundant eosinophils, patchy organization, fibrinous exudates, and interstitial thickening. One chronic EP case also showed focal LH. Additional features included intra-alveolar giant cells (6), pleuritis (3), small poorly-formed granulomas (3), and thrombi (2).

Conclusions: Lymphoid hyperplasia and eosinophilic pneumonia are previously unrecognized histopathologic manifestations of ALD, and amiodarone exposure should be included in their differential diagnosis.

P3743 Could interferon-gamma 1b have a role in treatment of fibrosing NSIP?
Barbara Bellolfiore, Marialuisa Bocchino, Giuseppe Antinolfi, Antonio Ponticiello, Roberta Di Grazia, Alessandro Sanduzzi Zamparelli, Sepideh Nikfam.
Internal Medicine, Division of Pulmonary Medicine Infection and Oncology, Nippon Medical School, Tokyo, Japan

Aim: To detect the effectiveness of Interferon-gamma 1b in patients with fibrotic NSIP.

Methods: We reviewed clinical records and pathologic findings of 11 patients with IFN (all males, mean age 54 ±6.3 yrs), who underwent surgical biopsy and were treated with IFN-gamma 1b between 2005 and 2007. In all patients, before therapy, after 6 and 12 (T2) months of treatment, the following parameters had been measured: lung volumes, DLCO, mPAP estimated by echodoppler. Biopsy slides were reviewed by two pathologists and reclassified according to the ATS/EIRS consensus classification of interstitial pneumonia.

Results: 5 out of 11 patients, previously diagnosed as Usual Interstitial Pneumonia, after the revision were identified as fibrotic NSIP. In Iapal UIP subgroup, 2 patients died after 6 months of treatment, while all the 5 patients with NSIP achieved at least 18 months of treatment with IFN gamma. Although the baseline values in NSIP group were better than those of UIP group, no significant difference in baseline lung function was observed between the two cohorts. In NSIP patients PV (70.16±18.5% at T0 vs 72.4±24.8% at T2, p =0.05), TLC (64.2±8.7% at T0 vs 67.7±14.8% at T2, p =0.05) and DLCO (47.3±17.2% at T0 vs 48.7±17.2% at T2, p =0.05) showed a slight improvement trend over time. Only the mPAP value showed a significant worsening after 12 months of treatment (27.18±6.8% at T0 vs 41.2±13.3% at T2, p =0.02).

Conclusion: In this post-hoc analysis IFN-gamma 1b therapy showed a trend toward reduced disease progression in NSIP patients. The effects on the deterioration of the mPAP values need further investigation through more invasive procedures, such as the right heart catheterization.
P3746

Combined pulmonary fibrosis and emphysema: Descriptive analysis from a specialized clinic of interstitial lung disease

M. Assunção Nieto1, Beatriz Morais1, Covadonga Fernandez-Golfin2, Paola Benedetti3, Gema Rodriguez-Trigo1, Iña Gerasimova1, José Luís Álvarez-Sala Walther1, 4, Pneumology, Hospital Clínico San Carlos: Universidad Complutense de Madrid, Madrid, Spain; 2Cardiology, Hospital Clínico San Carlos: Universidad Complutense de Madrid, Madrid, Spain

The combination of pulmonary fibrosis and emphysema (CPFE) is a recently defined syndrome characterized with pulmonary fibrosis (PF) and emphysema.

**Aim:** To analyze the clinical, functional, radiological and echocardiographic features in patients with CPFE.

**Method:** Prospective study conducted in a specialized clinic of ILD from 01/01/2007 to 31/12/2010. PF was defined as reticulonodular opacities with peripheral and basal predominance, honeycombing, architectural distortion and/or traction bronchiectasis without ground glass opacities in HRCT and emphysema as the presence of areas of low attenuation with a very thin or no wall and/or bullae with upper zone predominance.

**Results:** We anlized 20 patients, 90% men; mean age 74 years; 91% were smokers or ex-smokers, 60% had dyspnea, 75% basal cracks and 25% finger clubbing. The pulmonary function tests are shown in table 1. Pulmonary hypertension was detected in 5 of 10 patients who underwent echocardiography.

**Pulmonary function tests in 20 patients**

<table>
<thead>
<tr>
<th>Test</th>
<th>Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1/FVC %</td>
<td>72 (10)</td>
</tr>
<tr>
<td>FEV1% pred</td>
<td>86 (18)</td>
</tr>
<tr>
<td>FVC % pred</td>
<td>90 (17)</td>
</tr>
<tr>
<td>TLC %</td>
<td>81 (17)</td>
</tr>
<tr>
<td>RV % pred</td>
<td>88 (35)</td>
</tr>
<tr>
<td>RV/TLC % pred</td>
<td>104 (24)</td>
</tr>
<tr>
<td>TLCO % pred</td>
<td>46 (20)</td>
</tr>
<tr>
<td>PaO2 at rest</td>
<td>62 (9)</td>
</tr>
<tr>
<td>6 Minute Walking Distance</td>
<td>435 (96)</td>
</tr>
<tr>
<td>SP02 at exercise</td>
<td>84 (8)</td>
</tr>
</tbody>
</table>


**Conclusions:** The finding of lung volumes normal or minimally altered in contrast to a severe reduction in TLCO and arterial hypoxemia should alert to the possibility of CPFE. The prevalence of PH in these patients is high.

P3747

The King’s sarcoid questionnaire (KSQ): The development of a novel health related quality of life (HRQOL) questionnaire

Amir Patel1, Richard Siegenthaler1, Akim Sovremimo1, Daniel Creamer3, Genevieve Larkin1, Athol Wells1, Irene Higginson2, Surinder Birring1.

1Dept. of Opthalmology, King’s College Hospital, London, United Kingdom; 2Dept. of Palliative Care, Policy & Rehabilitation, Cicely Saunders Institute, King’s College London, London, United Kingdom; 3Dept. of Respiratory Medicine, Royal Brompton Hospital, London, United Kingdom; 4Dept. of Dermatology, King’s College Hospital, London, United Kingdom; 5Dept. of Ophthalmology, King’s College Hospital, London, United Kingdom

The preliminary King’s Sarcoid Questionnaire (KSQ) was tested in 205 patients (184 lung, 55 skin, and 45 eye disease) at two sites (King’s College Hospital and the Royal Brompton Hospital). The following items were removed: 1) floor effect >60% of participants responded “rarely” or “never” (8 items), 2) inter-item correlations of r<0.8 (12 items), 3) items that did not fit unidimensional scales following Rasch analysis (11 items). The King’s Sarcoid Questionnaire (KSQ) comprises of 5 modules: general QOL (12 items), lung (8 items), medication and side-effects (3 items), skin (4 items) and eyes (7 items). The KSQ is currently undergoing evaluation of test-re-test reliability and responsiveness to change.

P3748

Evaluation of the COPD assessment test (CAT) for measuring health-related quality of life in patients with interstitial lung disease

Kazuma Nagata, Keisuke Tomii, Michio Hayashi, Kojoro Otuka, Ryo Tachikawa, Kyoko Otsuka. Respiratory Medicine, Kobe City Medical Center General Hospital, Kobe, Japan

There is a need for a short, simple, and well validated instrument to assess health-related quality of life (HRQOL) in patients with interstitial lung disease (ILD). The study was conducted to evaluate the validity of the COPD assessment test (CAT), which is a newly developed short and simple questionnaire for COPD with good measurement properties, in ILD.

52 ILD patients completed the CAT and the St. George’s Respiratory Questionnaire (SGRQ). The patients also completed the MRC dyspnea scale, the Leicester Cough Questionnaire (LCQ), and the Hospital Anxiety and Depression Scale (HADS) and also underwent a six-minute walk test, pulmonary function tests, and arterial blood gas analysis.

The correlation between the CAT score and the SGRQ total score was very strong (r=0.93, p<0.0001). The CAT score was also significantly correlated with the SGRQ symptoms score (r=0.74, p<0.0001), with the SGRQ activity score (r=0.87, p<0.0001), and with the SGRQ impact score (r=0.89, p<0.0001). Stepwise multiple regression analysis demonstrated that the MRC score and the LCQ score were the most strongly contributing factors to both the CAT score and the SGRQ total score. The CAT is a short and simple questionnaire for assessing ILD health status with good measurement properties.

P3749

Fungal colonization in interstitial lung diseases

Ilya Sivokozov1, Olga Lovachev1, Eugene Shmel2, Elena Larionova1.

1Endoscopy Dept, CTB RAMS, Moscow, Russian Federation; 2Granulomatosis Diseases Dept, CTB RAMS, Moscow, Russian Federation

**Background:** Many of interstitial lung disease are tend to impair the immunity, thus patients in this group have a higher risk of fungal diseases. But still little is known regarding the role of fungal colonization in interstitial lung diseases, when fungi are not definite a causative agent.

**Aim:** To investigate the incidence and effects of fungal colonization of lower respiratory tract using PCR of bronchoalveolar lavage (BAL) specimens among patients with sarcoidosis and extrinsic allergic alveolitis (EAA).

**Materials:** 132 patients with sarcoidosis, and 80 patients with EAA underwent BAL with further PCR test for A. fumigatus and C. albicans using commercial assays. Cytology of BAL, different disease parameters including functional tests, radiological were analyzed.

**Results:** In sarcoidosis patients, Aspergillus was found in 22 cases (16.7%), 14 were males, and Candida detected in 11 cases (8.3%) - 3 were males. Among EAA patients, Aspergillus detected in 13 cases (16.2%), 5 were males, and Candida in 9 (11.3%) cases, 4 were males. In sarcoidosis, Candida colonization was associated with lower FVC (72%), DLCO (64%), and higher neutrophils level in BAL (32%), as well as disease duration (112.8 weeks) vs patients without it (92%, 88%, 9% and 38.2 weeks, respectively). In EAA Candida detection was also associated with lower DLCO (58%), 6-minute walk test distance (340 m), higher neutrophils level in BAL (34%), disease duration (107.3 weeks) vs patients without it (72%, 421 m, 12% and 48.7 weeks, respectively). Aspergillus detection altered disease parameters in similar way.

**Conclusion:** Colonization of respiratory tract with fungi in interstitial lung diseases plays a prominent role in deterioration of disease.

P3750

Forced vital capacity decreases rapidly in patients with idiopathic upper lobe-dominant pulmonary fibrosis

Kentaro Watanabe1, Taisi Harada1, Takako Hirota1, Kazuki Nabeshima2,

1Respiratory Medicine, Fukuoka University School of Medicine, Fukuoka, Japan; 2Pathology, Fukuoka University School of Medicine, Fukuoka, Japan

We present five patients with upper lobe-dominant pulmonary fibrosis of unknown etiology that does not fit any of the currently defined subsets of idiopathic interstitial pneumonias. We describe the clinical, functional, and pathological characteristics of this disorder, which we have provisionally termed idiopathic upper lobe-dominant pulmonary fibrosis (uULPF).

All patients were slender, with a body mass index of 16.0–17.9 kg/m². Four of the five patients had recurrent pneumothorax. Their pathological characteristics were as follows: 1) upper lobe-dominant subpleural proliferation of elastic fibers associated with deposition of mature collagen in alveolar lumens; 2) isolated fibrotic areas containing alveolar lumens filled with mature collagen were occasionally found in the lung parenchyma distant from the subpleural fibroelastosis; 3) visceral pleura adjacent to the subpleural fibroelastosis were often thickened and contained hyalinized collagen fibers; 4) there was an abrupt transition from subpleural fibrotic areas to less-involved pulmonary parenchyma; and 5) destruction of the lung architecture was minimal and fibroblastic foci were rarely seen. Ventilatory impairment was also characteristic of this condition. The median yearly decline in forced vital capacity in uULPF patients was -20.3% (range, -7.7% to -27.1%), which was more rapid than that reported for chronic fibrosing interstitial pneumonias such as idiopathic pulmonary fibrosis (IPF). uULPF is a unique pulmonary fibrosis that results in rapid deterioration of ventilatory function. It should be differentiated from other idiopathic fibrosing interstitial pneumonias.

676s

Abstract printing supported by G Chiesi. Visit Chiesi at Stand D.30
P3751 Pulmonary hypertension and right ventricular impairment in patients with interstitial pneumonia
Nina Karoli, Andrey Brevor. Hospital Therapy Department, Saratov State Medical University, Saratov, Russian Federation
There is little data about frequency of pulmonary hypertension (PH) and right ventricular (RV) impairment in patients with idiopathic interstitial pneumonia (IIP). In this work we examined the prevalence PH in these disorders using noninvasive cardiovascular evaluation. We studied 40 patients (aged 50.3±2.55) with IIP have been studied in early period after IIP diagnosed. Criteria of exception were the clinical displays heart disease, arterial hypertension. Control group have made 14 normotensive volunteers (aged 47.3±1.86). All patients were studied using realtime, phased array, two-dimensional Doppler echocardiography. The pulmonary artery systolic pressure (SP) was calculated as the sum of the tricuspid pressure gradient and the right atrial pressure. Results: Pulmonary hypertension (SP>35 mm Hg) is revealed at 15 patients with IIP and SP>40 mm Hg – at 7 (17.5%). Doppler-estimated SP at the patients with IIP exceed SP at the healthy persons only 4.9±1.1 mm Hg and 25.6±1.18 mm Hg, p<0.001. RV hypertrophy was present in 23 (57.5%) patients (p<0.001), RV dilation was present in 76.5% patients with IIP (p<0.001). RV diastolic dysfunction was revealed in 65% patients. Right atrial diameter in patients with IIP was significantly higher than at the healthy persons (p<0.001). RV wall in patients with IIP was significantly higher than at the healthy persons (p<0.001). RV diastolic dysfunction was revealed in 56% pts. We noted correlations between SP and dyspnoe intensity (scale MRS) (r=0.63, p<0.01), VC (r=0.61, p<0.01), FVC (r=0.39, p<0.01), FEVI (r=0.38, p<0.01), SaO2 (r=0.58, p<0.01).
Conclusion: Pulmonary hypertension and right ventricular abnormalities are common in patients with IIP.

P3752 Interstitial lung diseases in Europe
Roberto Carbone1, Rosangela Filiberti2, Edoardo Savarino1, Riccardo Ghio 1, P3752
Conclusion: SaO2 (r=-0.58, p<0.001) and SP (r=0.63, p<0.001) significantly higher than at the healthy persons (p<0.001). RV wall in patients with IIP was significantly higher than at the healthy persons (p<0.001). RV diastolic function was revealed in 56% pts. We noted correlations between SaO2 and SP and dyspnoe intensity (scale MRS) (r=0.63, p<0.01), VC (r=0.61, p<0.01), FVC (r=0.39, p<0.01), FEVI (r=0.38, p<0.01), SaO2 (r=0.58, p<0.01).

P3753 Effect of PDE-5 inhibitor treatment in patients with interstitial lung disease and pulmonary hypertension
Gregor S. Zimmermann1, Katharina Jakob1, Werner von Wulffen1, Werner Baumgartner2, Claus Neurohr 1.
Aim: To quantify the burden of the ILD in Europe. Methods: English articles on ILD epidemiology in Europe were reviewed using PubMed as the search engine. Results and conclusions: In Europe, ILD incidence rates ranged from 3.62 (South of Spain) to 7.6 cases ×100,000 inhabitants (Spain) according to different studies. In the majority of cases a male predominance was found. Among ILD, the most frequent entities were idiopathic pulmonary fibrosis (IPF: 19-39% of all ILD), sarcoidosis (12-35%) and hypereosinophilic pneumonia (3-12%). Incidence rates of IPF and sarcoidosis ranged from 0.93 (Greece) to 6.78 cases ×100,000 (UK) and from 1.07 (Greece) to 5.59 ×100,000 (UK), respectively. Median age at diagnosis was about 61 years for IPF and 42 years for sarcoidosis. Fibrosis NSIP was considered as a new subgroup, making up 20-35% of patients previously diagnosed as IPF. Few data exist on the frequency of ILD and discrepancies among different areas have been observed. Differences in part real, but are due to the use of inconsistent diagnostic sources (diagnostic registries, selected clinical series, different study designs), to the inclusion of incident or prevalent cases, to variable criteria for diagnosis, or to selection bias.

P3754 Pulmonary hypertension in patients with interstitial lung disease
Zeynep Dogum1, Sinan Erginel1, Huseyin Yildirim1, Guntulu Ak1, Ragip Ozkan2, Fusun Alatas1, Musaffer Metinbas1. 1Department of Chest Disease, Eskisehir Osmangazi University, Medical Faculty, Eskisehir, Turkey; 2Department of Radiology, Eskisehir Osmangazi University, Medical Faculty, Eskisehir, Turkey
Pulmonary hypertension (PH) is commonly seen in patients with interstitial lung disease (ILD), and is associated with a worse prognosis. The aim of this study was to determine the prevalence and characteristics of PH in patients with ILD. Demographic and clinical characteristics, physiological studies, six-minute-walking test (6MWT) and high resolution computed tomography (HRCT) results were prospectively collected, and compared between patients with and without PH. Pulmonary hypertension was defined by right heart catheterization as mean pulmonary artery pressure > or =25 mm Hg and pulmonary occlusion pressure < or =15 mm Hg. The study cohort consisted of 30 patients. The final diagnoses of these patients were idiopathic pulmonary fibrosis (IPF: 19 patients), asbestos-related IPF (5), connective tissue disease-associated IPF (5), chronic hypersensitivity pneumonitis (n=3), and un-classified (n=4). Fourteen patients (46.6%) were found to have PH. The mean pulmonary arterial pressure in PH patients was 32.9±6.8 mm Hg, and 18.2±3.5 mm Hg in non-PH patients (p<0.001). When compared with non-PH patients, subjects with PH exhibited lower six-minute-walk-distance (415±41 m vs. 260±19 m, p<0.001). In addition to these, patients with PH had the following clinical characteristics: advanced HCT fibrosis score, decreased oxygen saturation during 6MWT, and decreased percentage of predicted FVC, percentage of predicted FEV1, and percentage of predicted diffusing capacity for carbon monoxide. Our findings revealed that pulmonary hypertension occurs commonly in ILD. Pulmonary hypertension plays an important and underappreciated role in the functional status of patients with ILD.

P3755 Resting PaO2 and 6MWT as diagnostic index for nocturnal oxygen desaturation in diffuse parenchymal lung diseases
Sheenu Singh1, Manoj R Lal Gupta2, Ravinder Singh2, Virendra Singh2. 1 Chest & Tuberculosis, SMS Medical College, Jaipur, Rajasthan, India; 2 Pulmonary Medicine, SMS Medical College & Hospital, Jaipur, Rajasthan, India
Introduction: Despite of normal daytime oxygen saturation many patients of diffuse parenchymal lung disease (DPLD) desaturate during night time. Such patients may develop signs of pulmonary arterial hypertension (PAH) which increases the morbidity and mortality in DPLD. Our aim was to do the magnitude of nocturnal oxygen desaturation and find its predictors in DPLD. Methods: It was a cross sectional and observational study. 48 consecutive patients of DPLD were recruited. Idiopathic pulmonary fibrosis (IPF) was diagnosed by ATS/ERS guidelines 2002 and other DPLD by their clinical and radiological presentation. Arterial blood gas analysis, six minute walk test (6MWT) and overnight pulse oximetry were done and variables such as time spent with nocturnal oxygen saturation (SpO2) below 90%, mean SpO2, worst SpO2 and apnea - hypopnea index were recorded. Results: Amongst 48 subjects, 35 (72.9%) patients had IPF, 9 (18.8%) patients had hypersensitivity pneumonitis and 4 (8.3%) patients had stage 4 sarcoidosis. Nocturnal desaturation (>10% of sleep time with SpO2<90%) was observed in 20 (41.7%) subjects. The desaturators were found to have resting daytime PaO2 of less than 60mm of Hg (p=0.0015), end SpO2 after 6MWT <83.5% (p=0.0077) and distance walked during 6MWT ≤380.4m (p=0.0051). Based on cut off values of these variables an index called desaturation index was evolved which had 95% sensitivity. Conclusion: Significant nocturnal oxygen desaturation occurs in many patients of DPLD despite of normal resting SpO2. A desaturation index based on PaO2 and end SpO2 after 6MWT and distance walked during 6MWT was found to be 95% specific in diagnosing nocturnal desaturation.
P3756
Drug induced lung disease: 11 cases
Eldy Kopeli, Tigge Sahin Odemislet, Zeynep Erzyaman Ozen, Gaye Ulubay, Sule Akcaay, Fusun Oner Eynoboglu. Pulmonary Diseases, Baskent University School of Medicine, Ankara, Turkey

Drug induced lung disease (DILD) is the most common cause of iatrogenic damage to the lungs. More than 350 drugs have been identified leading to DILD. We evaluated the characteristics of patients with DILD between 2007-2010, at our institution retrospectively, for demographic features, pulmonary function tests (PFTs), diffusion capacities, high resolution computed tomograms (HRCT) findings, diagnostic methods and treatment modalities. 11 patients (M=9,2, mean age 60±20.3) were identified with DILD. 3 had chronic myeloid leukemia; congestive heart failure, asthma, testicular cancer, malignant melanoma, acute lymphocytic and myelocytic leukemia, renal transplantation and lung cancer was diagnosed in one patient each. Cough, respiratory failure and fever were the most common symptoms. 4 patients exhibited normal, 5 revealed restrictive, one patient for each case obstructive and mixed PFTs. Diffusion capacities in 6 patients were low. Ground glass opacities (2), bilateral patchy consolidation (2), interstitial fibrosis (2), pleural effusion (1) were evident in HRCT, however 1 patient revealed no abnormality. 3 patients underwent bronchoscopy for bronchoalveolar lavage. Infectious causes were excluded by microbiological and laboratory tests. Lung involvement was thought to be due to bleomycine (1), methotrexate (1), cyclophosphamide (1), cisplatin (1), dasatinib (2), ARA-C (2), cilazapril (1), amiodaron (1), tacrolimus (1). Radiological and clinical improvement was achieved with systemic steroids in 10 and cessation of the drug in one. DILD can cause significant mortality and morbidity. We believe that consequence of DILD could be reduced by high degrees of suspicion and by excluding other causes for common pulmonary symptoms and abnormal radiographic findings.

P3757
Quadriiceps function is reduced in fibrotic idiopathic interstitial pneumonia
Laura Mendoza1, Athena Gogali 2, Samuel Kemp3, Dinesh Shrikrishna 3, Toru Arai1,2, Yoshikazu Inoue2 , Kazunobu Tachibana1,2, Yasushi Inoue1, Maria Ntouskou4 , Anastasios Kalliamo 5, Oramia Anagnostopoulou 1, George Tezlepil 1, 25th Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 22nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 1st Cardiology Clinic, University of Athens, Athens, Greece; 22nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 2nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 1st Cardiology Clinic, University of Athens, Athens, Greece; 22nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 2nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 2nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 2nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; 2nd Pulmonary Clinic, General Hospital of Chest Diseases “Sotiria”, Athens, Greece; Boston Bantestrak Medical Center, University of Athens, Athens, Greece; 1st Clinic of Pathophysiology, University of Athens, Athens, Greece

Background: Cardiac involvement remains an important prognostic factor in patients with sarcoidosis. However, early diagnosis of cardiac sarcoidosis has been difficult because the clinical manifestations are not specific and the sensitivity and specificity of the diagnostic modalities are limited.

Methods: Patients with biopsy proven sarcoidosis were prospectively recruited from the outpatient clinic of sarcoidosis and underwent a full cardiopulmonary monitoring including pulmonary function tests, a 12-lead electrocardiography, echocardiography, a 24-hour ambulatory ECG and cardiac magnetic resonance imaging (MRI) when appropriate. Cardiac involvement was assessed based on known established Japanese Ministry of Health (JMH) criteria and on modified criteria using MRI as a major criterion for cardiac sarcoidosis. All consecutive patients were followed for 5 years for major adverse events.

Results: Seventy patients (43 female) were enrolled with median age 49.96±12.83 years old and disease duration 4.44±5.27 years. Cardiac involvement was identified with the modified criteria in 27 patients (38.5%) while 10 (14.3%) patients were found based on the JMH criteria, showing a more than two-fold higher rate for the MRI group (p=0.005). On follow-up, 9 patients had adverse events including 5 cardiac deaths. All patients with cardiac cause of death had cardiac sarcoidosis based on the modified criteria.

Conclusion: Evaluation of myocardial involvement in sarcoidosis with the use of MRI as a major criterion appears to be more sensitive than current consensus criteria and associated with future adverse events including cardiac death. Cardiac MRI evaluation may be of great importance in the early diagnosis of cardiac sarcoidosis.

P3759
Prognostic significance of serum markers in acute exacerbation of idiopathic interstitial pneumonias
Tora Arai1,2, Yoshikazu Inoue2, Kazunobu Tachibana1,2, Yasushi Inoue1, Akiko Ishii Kishiyama1, Chikatso Sogumoto1, Tomoko Kagawa2, Toshinobu Okuma1, Masanori Akira1, MARott Kitaichi1, Seiji Hayashi1, 1Department of Respiratory Medicine, National Hospital Organization Kind-Kuicho Chest Medical Center, Sakai, Osaka, Japan; 2Clinical Research Center, National Hospital Organization Kind-Kuicho Chest Medical Center, Sakai, Osaka, Japan; 4Department of Radiology, National Hospital Organization Kind-Kuicho Chest Medical Center, Sakai, Osaka, Japan; 5Department of Pathology, National Hospital Organization Kind-Kuicho Chest Medical Center, Sakai, Osaka, Japan

Introduction: Acute exacerbation (AE) of idiopathic interstitial pneumonias (IIPs) is a topic for recent years. We have shown that diffuse pattern of high resolution CT (HRCT) at AE of idiopathic pulmonary fibrosis (IPF) is a worse prognostic factor (Akira, AJRCCM 2008). Prognostic significance of serum levels of KL-6 and surfactant protein (SP)-D in AE of IIPs has not been clarified sufficiently.

Aim: We examined clinical findings in AE of IIPs retrospectively to clarify the significance of serum KL-6 and SP-D as a prognostic factor.

Methods: Serum levels of KL-6 and SP-D were compared between 30 days survivors and non-survivors. Prognostic factors determining 30 days survival was examined by logistic regression analysis among various clinical parameters including serum markers, HRCT patterns at AE (diffuse/non-diffuse).

Results: Serum KL-6 levels of 30 days survivors at AE were significantly less than that of non-survivors, however, there was no difference in serum KL-6 levels at AE. Serum KL-6 of patients with diffuse pattern was significantly lower than that of patients with non-diffuse pattern. Logistic analysis revealed that serum KL-6 at AE, white blood cell counts, immunoglobulin G, gender, complication of diabetes mellitus and pre-treatment before AE were significant factors determining 30 days survival.

Conclusions: Serum KL-6 (≥1450U/ml) is a significant worse prognostic factor determining 30 days survival in AE of IIPs.