389. Diffuse parenchymal lung disease III

P3650
The decrease of surfactant protein D in bronchoalveolar lavage fluid in patients with idiopathic pulmonary fibrosis and nonspecific interstitial pneumonia
Hirotaka Nishikiori, Hirofumi Chiba, Mitsuo Otsuka, Koji Kuronuma, Hiroki Takahashi. Third Department of Internal Medicine, Sapporo Medical University School of Medicine, Sapporo, Hokkaido, Japan

Background: Surfactant protein (SP)-A and SP-D are useful biomarkers for the diagnosis and evaluation of activity of interstitial lung diseases. SP-A and SP-D, which are lung specific proteins, belong to a subgroup of the C-type lectin superfamily. It has been reported that these proteins play important roles in fibrotic lung. Although they are mainly produced by type II pneumocytes and Clara cells, it is not clarified these production and clearance in fibrotic lung.

Methods: To elucidate those issues, we measured levels of SP-A, SP-D and KL-6 by enzyme-linked immunosorbent assay in BAL fluid (BALF) and serum of 24 IPF patients, 36 NSIP patients (8 patients diagnosed by surgical biopsy, 28 patients clinically diagnosed) and 17 sarcoidosis patients. The levels of SP-A and SP-D in BALF were compared with those from 20 healthy controls. We investigated also the relationship of protein levels between serum and BALF.

Results: In IPF and NSIP patients, SP-D levels in BALF were significantly lower than those from healthy controls (p=0.006 and p=0.003) and sarcoidosis patients (p=0.02 and p=0.01). SP-A levels in BALF were no significant difference among these patients and controls. The significant positive correlation of SP-D levels between serum and BALF was found in IPF patients (r=0.529, p=0.008). In NSIP patients, the correlation of SP-D levels between them was not significant. No correlation of SP-A levels between serum and BALF was observed in any patients groups.

Conclusion: In IPF patients, SP-D levels in BALF were lower than those in healthy controls and had significant positive correlation with those in serum.

P3651
Characteristics of inspiratory and expiratory reactance in interstitial lung disease
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Forced oscillometry is a noninvasive method to measure respiratory impedance and widely used in obstructive lung diseases, but has not been well studied in restrictive lung diseases. This study was conducted to investigate the characteristics of measurements obtained by impulse oscillation system (IOS) in patients with interstitial lung disease (ILD).

IOS and spirometry were performed in 64 ILD patients, 54 asthma patients, 49 chronic obstructive pulmonary disease (COPD) patients, and 29 controls. Respiratory resistance and reactance were assessed as measurements averaged over several tidal breaths (whole-breath analysis) and as measurements separately averaged during inspiration and expiration (inspiratory-expiratory analysis). Whole-breath analysis failed to distinguish between ILD and obstructive lung diseases. Inspiratory-expiratory analysis demonstrated no difference between inspiratory and expiratory reactance at 5 Hz (X5) in controls and asthma patients. Expiratory X5 was more negative than inspiratory X5 in COPD patients. In contrast, inspiratory X5 was found to be less negative than inspiratory X5 in ILD patients. Furthermore, within-breath change in X5 was inversely correlated with vital capacity and diffusing capacity of carbon monoxide in ILD patients. These results suggest that increased magnitude of X5 during inspiration compared with that during expiration is a characteristic feature of IOS measurements in ILD patients.
The efficacy of pirfenidone in scleroderma related interstitial lung disease (SSc-ILD)

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Introduction: The major cause of death in Systemic Sclerosis (SSc) is interstitial lung disease, and cyclophosphamide is an only agent which significantly demonstrated a beneficial effect on lung function in patients with scleroderma-related interstitial lung disease (SSc-ILD), however the effect was quite modest, and it is necessary to identify a reasonable alternative.

Objectives: TGF-beta1 plays a critical role in the pathophysiology of pulmonary fibrogenesis. Pirfenidone exerts its antifibrotic effect through regulation of TGF-beta1 levels. This raises the possibility that agents targeting TGF-beta1 may be beneficial for SSc-ILD.

Methods: We administered pirfenidone to 3 patients with SSc-ILD and evaluated pulmonary function.

Results: Case 1 is a 62 year-old female. Vital capacity (VC) improved by pirfenidone. The change rate was +27.3% (+0.51L) for 5 months. Case 2 is a 75 year-old female. VC improved remarkably, at the change rate of +44.4% (+0.32L) for 25 months. Case 3 is a 66 year-old female. VC improved at the rate of +8.3% (+0.17L) for 26 months.

Conclusion: All of 3 patients with SSc-ILD demonstrated the favorable efficacy of VC by pirfenidone without severe adverse events. The previous studies documented that deteriorating lung function was associated with increased mortality in SSc-ILD. Therefore, it is necessary to identify and treat early stages of patients with SSc-ILD, preventing pulmonary function impairment. Pirfenidone exerts its antifibrotic effect through regulation of TGF-beta1, which is one of the important inducers of fibrogenesis in SSc. We suggest pirfenidone may be a possible option for SSc-ILD.

Effect of pirfenidone on chronic interstitial pneumonia

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Background: Several reports reveal the favorable effect of pirfenidone on early stage of idiopathic pulmonary fibrosis (IPF), but it is not clear on specific interstitial pneumonia (NSIP) and interstitial pneumonia (IP) associated with collagen-vascular diseases (IP-CVD).

Objectives: To examine the effect of pirfenidone on chronic IP including IPF, NSIP and IP-CVD.

Methods: Thirty-two patients were enrolled in the study evaluating the safety and efficacy of pirfenidone in IP, NSIP and IP-CVD. Clinical diagnosis are IPF (14), NSIP (14), IP associated with scleroderma (3), and rheumatoid arthritis (1). Based on PaO2 at rest and SpO2 after 6 minutes walk test (6MWT), disease severity of those were classified into four groups [Table 1]. We retrospectively analyzed subjective symptom in British Medical Research Council scale, pulmonary function, KL-6, SP-D and CT findings before and after pirfenidone administration.

Table 1. Disease severity of patients

<table>
<thead>
<tr>
<th>PaO2 at rest (mmHg)</th>
<th>SpO2 after 6MWT</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>&gt;80</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>70-79</td>
<td>When &lt;90%, to III</td>
</tr>
<tr>
<td>III</td>
<td>60-69</td>
<td>When &lt;90%, to IV</td>
</tr>
<tr>
<td>IV</td>
<td>&lt;60</td>
<td>unnecessary to perform 6MWT</td>
</tr>
</tbody>
</table>

Results: Correlations was observed between subjective symptom and VC, but was not between subjective symptom and KL-6, SP-D, CT findings. After administration of pirfenidone, VC improved above 5% in 22%, 50%, 25%, 45% of patients (severity grade I, II, III, IV, respectively). Subjective symptoms highly improved in patients with scleroderma (improvement ratio: 67% in scleroderma vs. 14%, 15% in IPF, NSIP, RA, respectively).

Conclusions: Pirfenidone might have the favorable efficacy independent of disease severity, without severe adverse events, in chronic IP, especially in IP associated with scleroderma.

Telephone consultations for follow-up of interstitial lung disease: Patient satisfaction survey

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Introduction: The long-term follow-up required by Interstitial Lung Disease (ILD) patients imposes a significant burden on healthcare resources. Telephone consultations have been piloted as an alternative to some face-to-face consultations in a teaching hospital based ILD service. Patients whose next consult is anticipated to be uncomplicated (eg stable patients having interval lung function tests) are offered a phone consult with the ILD consultant.

Aims and objectives: To assess whether ILD patients are as satisfied with a telephone consultation as with a face-to-face consult.

Methods: A postal survey was sent to 95 consecutive ILD patients who had a phone consult over a 12-month period. Patients used a 5-point score to report if in comparison to a face-to-face consult, the phone consult was as punctual, convenient, reassuring, and afforded the same opportunity to ask questions, and facilitated the same understanding of information.

Results: 52 patients (55%) posted responses (see Table 1). 67% agreed phone consults were as punctual, 78% agreed they were as convenient, 77% felt as reassured and as able to ask questions. 88% understood the information given just as easily. 80% were willing to have another phone consult.

Table 1

<table>
<thead>
<tr>
<th>%</th>
<th>Strongly Disagree</th>
<th>Disagree</th>
<th>Neutral</th>
<th>Agree</th>
<th>Strongly No</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>As punctual</td>
<td>2</td>
<td>17</td>
<td>12</td>
<td>40</td>
<td>27</td>
<td>2</td>
</tr>
<tr>
<td>As convenient</td>
<td>12</td>
<td>30</td>
<td>11</td>
<td>48</td>
<td>30</td>
<td>3</td>
</tr>
<tr>
<td>Felt as reassured</td>
<td>2</td>
<td>10</td>
<td>11</td>
<td>48</td>
<td>29</td>
<td>0</td>
</tr>
<tr>
<td>Able to ask questions</td>
<td>0</td>
<td>11</td>
<td>12</td>
<td>44</td>
<td>33</td>
<td>0</td>
</tr>
<tr>
<td>Understood the information</td>
<td>0</td>
<td>6</td>
<td>6</td>
<td>52</td>
<td>36</td>
<td>0</td>
</tr>
</tbody>
</table>

Discussion: The majority of ILD patients selected for a telephone consultation were as satisfied as with a face-to-face consult. Telephone consultations are acceptable to the majority of ILD patients for at least some of their follow-up.

NSIP: A diagnosis?

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Introduction: Non-specific interstitial pneumonia (NSIP) is the most discussed entity in the group of idiopathic interstitial pneumonias. Recognized in 2002 by the American Thoracic Society and the European Respiratory Society as a provisional diagnosis, NSIP may be idiopathic or associated with a number of clinical settings.

Objective: The objective of this study was to determine the proportion of strictly idiopathic NSIP among a cohort of patients whose initial lung CT scan considered NSIP as a possible diagnosis.

Method: 73 adult patients with an initial CT pattern of NSIP were selected in our database of adult patients with ILD (1998-2011) and were enrolled in a monocentric retrospective study. Anamnestic, clinical, functional, biological and pathological data were reviewed for each patient as well as the evolution of disease.

Results: In only 21 patients of 73 (28.9%), a definitive diagnosis of idiopathic NSIP could be retained. In 10 of 21, an open lung biopsy was performed and showed cellular NSIP in 2, and fibrous NSIP in 8. In the remaining 52 patients (71.2%) an alternative diagnosis could be made, most frequently an underlying connective tissue disease (n=22, 30.1%) and idiopathic pulmonary fibrosis (n=12, 16.4%).

The other diagnoses include hypersensitivity pneumonitis (n=7, 9.6%), drug-induced lung disease (n=4, 5.5%), sarcoidosis (n=4, 5.4%), a sequel of ARDS (n=1, 1.4%), LIP (n=1, 1.4%) and a RBILD (n=1, 1.4%).

Conclusion: This study demonstrates that idiopathic NSIP is a rare condition among patients presenting a radiological NSIP pattern. Therefore an integrated multidisciplinary approach is recommended in order to diagnose alternative/associated diseases needing for most of them a specific treatment.
In the analysis of 6MWD data from a large cohort of IPF patients yielded an MCID of 22–37 m. This finding is consistent with previous estimates which, taken together, provide a meaningful benchmark for assessing 6MWD in patients with IPF.

P3657
The efficacy of transbronchial biopsies without fluoroscopy control in diagnostics of extrinsic allergic alveolitis
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Background: Extrinsic allergic alveolitis (EAA) is usually established based on results of CT exam, immunology, and transbronchial lung biopsy (TBLB) during flexible bronchoscopy (FBS). As a rule, TBLB should be performed under fluoroscopy control, but it is not always possible, and in such situation efficacy of TBLB is controversial.

Aim: To compare the effectiveness of TBLB and bronchoalveolar lavage (BAL) during FBS in patients with newly diagnosed EAA.

Materials: 66 patients with EAA were enrolled into the study, all of them underwent CT of the chest with subsequent FBS with TBLB and BAL. We analyzed the diagnostic efficacy of TBLB upon histology and cytology, and diagnostic changes in BAL. Additionally, mean and median number of tissue samples, samples quality, frequency of pneumothorax and severe bleeding were analyzed.

Results: There were 37 females in the group, mean age for both sexes - 48.3 years, varied from 19 to 73 years. Mean disease duration from first symptoms/chest abnormalities was 87.1 weeks. Mean tissue samples number was 2.23, varied from 1 to 5, median 2. There were no complications. Lung tissue was observed in 4266 cases (64.5%). Upon histology, granulomas and tissue infiltration were seen in 30 out of 66 patients (45.4%), additionally due to cytology granulomas were found in 4 cases, thus total efficacy of TBLB was 34/66 (51.5%). Among the rest 32 patients, diagnostically significant changes in BAL were found in 16 patients, so the total efficacy of TBLB plus BAL reached 50/66 (75.7%).

Conclusions: Bronchoscopy with TBLB and BAL even without fluoroscopy control is effective and safe way to establish the diagnosis of allergic alveolitis.

P3668
KL-6 compared to LDH as a prognostic factor in Caucasian patients with idiopathic pulmonary fibrosis
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Background: Medialinal sarcoidosis contributes a wide differential diagnosis, including sarcoidosis, tuberculosis, and malignancy. Endobronchial ultrasound-guided Transbronchial Nodule Aspiration (EBUS-TBNA) allows safe access to these nodes.

Methods: 119 patients with mediastinal lymphadenopathy were referred to our tertiary centre for EBUS-TBNA between January 2010 and August 2012. Final diagnoses were used to explore the utility of the 4 tests performed through EBUS-TBNA: immunology (IGRA and/or Tuberculin Skin Test), cytology; microscopy/culture; and the GeneXpert MTB/RIF integrated TB PCR assay.

Results: 27 patients (23% of cohort) were diagnosed with sarcoidosis based upon consistent clinical features and supportive TBNA. Cytology was predominantly non-caseating granulomata (24/27 (89%); 3/27 reactive). Immunology was negative in 13/22/5 (98%), positive in 3/25, not tested in two patients. Sensitivity and specificity for sarcoidosis with immunology alone was 87% and 69%, respectively, with specificity improved by cytology to 91%. GeneXpert was negative in all six further cases otherwise inconclusive by combined immunology/cytology testing (reactive appearances (3), immunology positive (3) or not done (2)). Three patients failed empirical TB treatment and were later re-diagnosed as sarcoidosis; all were PCR negative.

Conclusions: Combined immunology/TBNA/cytology data provide good specificity for sarcoidosis in patients with mediastinal lymphadenopathy. The inclusion of negative GeneXpert MTB/RIF in our cohort provided support in diagnosis for an additional 29% of cases. Quad-testing provides a fast-track and accurate diagnosis of sarcoidosis over TB.

P3660
Viral infection immunodetection reinforce the possible viral participation in the pulmonary fibrosis disease
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Background: Many patients with pulmonary fibrosis disease have been postulated an infectious etiology. In this regards, the aim of this study was investigate the presence or absence of virus infections in patients with pulmonary fibrosis disease.

Methods: The study sample comprised 38 patients with open lung biopsies, 13 patients with IPF/UIP; 6 with NSIP/UIP; 8 with NSIP/NSIP; 13 with AIP/DAD and 4 with ICLF/CLF.

The clinical/radiology and histological diagnosis was according to the 2001 ATS/ERS consensus. Micrarray samples were analyzing by immunohistochemistry for measles virus (MV), cytomegalovirus (CMV), hepatitis-C virus (HCV), adenosivus (ADV), respiratory syncytial virus (RSV), Epstein-Barr virus (EBV),
P3661 Pirfenidone, proton pump inhibitor, N acetyl cystine (PINPOINT) therapy for IPF: Tolerance and safety profile among Indian patients

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1Pulmonology, Lung Care & Sleep Centre, Vashi, Navi Mumbai, Maharashtra, India; 2Institute of Pulmonology, Medical Research & Development, Mumbai, Maharashtra, India

Rationale: To our knowledge there are no data of Pirfenidone, PPIs and N-acetyl cystine (PINPOINT) therapy for IPF in Indian patients. We retrospectively examined the safety & tolerability of PINPOINT in IPF-UIP.

Methods: 40 consecutive patients with IPF-UIP administered PINPOINT, Pirfenidone 200 mg three times day & titrated to 400 mg three times day over 2 to 4 weeks, a proton pump inhibitor & N-acetyl cysteine 1800 mg/day. Baseline functional lung tests were performed. Lung function & 6MWT were possible in 25 patients. Patients were followed for at 2 weeks, then monthly/quarterly. Prednisolone was added on follow up based on clinical assessment. Four newly diagnosed patients were given only PINPOINT.

Results: Baseline mean spO2 at rest was 95% & mean FVC 1.30 litres (55% predicted). Baseline liver function were normal in all patients. Mean lowest spO2 on 6 minute walk test was 90%. There was no significant increase in liver enzymes at follow up. 17 patients had pulmonary hypertension on 2D Echo. Mean duration of follow up was 241 days. In 25 patients, Pirfenidone could be increased to 1200 mg/day. Dose could not be increased to 1200 mg/day in 11 patients due to gastrointestinal side effects (nausea/vomiting 10 patients, loose motions - 1 patient). Pirfenidone was stopped in five patients because of skin itching & rash (4 patients 600mg/day, 1 patient 1200 mg/day), 5 patients continued to take Pirfenidone despite skin itching (no skin discoloration) after counseling about side effects & took symptomatic treatment. 5 patients expired during this period.

Conclusion: PINPOINT therapy appears to be well tolerated in patients with IPF-UIP.

P3662 Frequency and impact of interstitial lung disease on clinical state and mortality in systemic sclerosis

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1Internal Medicine, Tabet Sfar Hospital, Mahdia, Tunisia; 2Pneumology, Tabet Sfar Hospital, Mahdia, Tunisia; 3Internal Medicine, Fattouma Bourguiba Hospital, Monastir, Tunisia

Background: Interstitial lung disease (ILD) is a frequent complication and a leading cause of morbidity and mortality in Systemic Sclerosis (SSc). The aim of our study was to determine the frequency of ILD in a cohort of Tunisian SSc and to assess the influence of this association on clinical, biological features and outcome of the disease.

Methods: A retrospective study included 30 consecutive patients with SSc who fulfilled the American Rheumatism Association’s criteria. Patients were divided in two groups according to the occurrence (group 1) or not (group 2) of ILD. We compared the differences in the clinical, laboratory features and outcomes between the two groups. The data were compared by the Chi square corrected by fisher exact test and student test.

Results: Thirty patients (43, 3%) developed ILD. The patients were 12 women and one men, aged between 15 and 71 years (mean: 47.1 years) with the duration of the disease ranging from 4 to 348 months (mean: 81.7 months). The most common presenting complaints are dyspnea on exertion in 76.9% and dry cough in 46.1%. ILD was asymptomatic in 3 cases (23%). The comparison of the two groups showed that digital pitting, inflammatory synovitis and anti-Scl70 antibodies were significantly more common in the group 1. No differences were noted concerning sex ratio, mean age at SSc onset and delay of diagnosis. However, mortality rate was significantly higher in patients with ILD (46.1% vs 6.2%, p=0.025).

Conclusion: Our results confirm that ILD is a common manifestation of SSc, mainly encountered in patients with anti-SCL 70 antibodies and associated with poor prognosis.

P3663 Pirfenidone in idiopathic pulmonary fibrosis (IPF): Early single centre Irish experience

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Introduction: Pirfenidone is an orally bio-available synthetic molecule, which has recently been approved for the treatment of mild to moderate IPF in Europe. It regulates the activity of TGF-β and TNF-α in vitro. Open label pirfenidone prescription commenced in June 2011 in the Republic of Ireland. We report the early experience of a single centre with pirfenidone.

Methods: We conducted a retrospective review of medical records of those patients who were prescribed pirfenidone. We analysed baseline demographics, symptoms and pulmonary function. Comparisons between groups were conducted using paired t-testing.

Results: 26 patients (20 male) received the medication. 22 patients remain on the medication. 1 patient died due to an exacerbation of IPF and 3 others discontinuing the medication secondary to side effects. 15 (58%) of patients have reached target dose of medication. 7 subjects continue to take pirfenidone at a reduced dose. 14 participants reported side effects potentially related to pirfenidone. The most commonly reported side effect was fatigue followed by gastro-intestinal disturbance and photo-sensitivity. An increase (< 2 fold) in transaminases was noted in 1 patient. Patients who experienced side effects or required dose reduction were on average older but this did not reach statistical significance. Amongst subjects who had repeated pulmonary function testing (n=14), there was no significant decline in TLC or FVC between baseline and follow up.

Conclusion: Pirfenidone is a novel agent for the treatment of limited IPF. The side effect profile in an Irish population appears consistent with recent published data. Further follow up is required to establish efficacy in an Irish population.

P3664 UIP-like or NSIP pattern in interstitial lung disease patients (ILD), following by ‘connective tissue disease’ (CTD)

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1Pulmonary Department, Athens Chest Hospital 'Sotiria', Athens, Greece; 2Pathology Department, Athens Chest Hospital 'Sotiria', Athens, Greece; 3Radiology Department, Athens Chest Hospital 'Sotiria', Athens, Greece;

Aim: A retrospective study was performed on the follow up (FU) of ILD patients who developed CTD during at least 3 years follow up period.

Methods: A cohort of 120 ILD cases followed on clinico-pathological and HRCT findings. A number of 12 cases who were undertaken surgery biopsy (OLBIATS) were selected for the study.

Results: The above 12 patients were followed during the follow up period CTD cases have f-NSIP on the background and 6 have UIP-like pattern.

Conclusion: The ILD cases without clinical history of CTD and with UIP-like or NSIP pattern on the background of histology specimens, must be followed for long time period.

It is of important value such patients to be checked initially and in follow up by HRCT, clinical and laboratory tests.

In these subgroup of patients the prognosis is better than UIP/IPF patients.

P3665 Pulmonary functions in systemic sclerosis

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1Internal Medicine, Aleppo University Hospital, Aleppo, Syrian Arab Republic; 2Rheumatology, Aleppo Faculty of Medicine, Aleppo, Syrian Arab Republic

Background: Pulmonary involvement is the leading cause of systemic sclerosis (SSc)-related deaths.

Objective: To determine the prevalence of pulmonary involvement in systemic sclerosis and its distributions on the disease subsets, and to study its relation with other organ involvement.

Methods: We analyzed 43 SSc patients, a careful history and clinical examination was performed to each patient. All the patients were submitted to chest radiographs and high-resolution CT (HRCT) and underwent pulmonary function testing and echocardiography and electrocardiography and six minutes walk test. Patients were grouped into four descriptive disease subsets: lcSSc, dcSSc, SSc sine scleroderma and overlap-syndrome.

Results: Of the 43 patients, (53.5%) of patients had lcSSc and (27.9%) dcSSc. Overlap syndrome was diagnosed in (11.6%) of patients, SSc sine scleroderma was present in (7%) of patients, (93%) were female and the mean age at diagnosis was 41.67±11.51, (55.8%) of the patients had pulmonary symptoms or signs, dyspnea was the most prevalent symptom (37.2%). Pulmonary fibrosis was most prevalent involvement (41.9%), and it was more common in dcSSc (41.6%) compared with lcSSc (30.4%). Pulmonary hypertension was second in prevalence (PASP>35 mmHg) 20.9% and it was more common in lcSSc (21.7%) compared with dcSSc (8.3%). (62.8%) of the patients had restrictive pulmonary function test. (65.1%) of the patients had normal six minutes walk test.

Discussion: Our results will be discussed with well known international studies.
P3668
Respiratory bronchiolitis interstitial lung disease – What is the natural history?
Andrew Long1, Jason Viner1, Anthony Edey2, Ann Millar1,3.

The natural history of RBILD remains uncertain with contradictory views based on current data. In order to investigate the effect of smoking cessation on the long term physiological and radiological outcome of RBILD we have retrospectively identified patients with this diagnosis between 2003-2011 (CT and pathological diagnosis). 12 patients with biopsies consistent with RBILD were included with a median 36 pack year history. 4 patients successfully quit smoking (Quitters) while 8 either continued to smoke or were inconsistent in their cessation attempts (Smokers). Baseline lung function at diagnosis was compared to current values of FEV1, FVC, TLCO and KCO between Quitters and Smokers:

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Smokers (n=8)</th>
<th>Quitters (n=4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1</td>
<td>Improved</td>
<td>Declined</td>
</tr>
<tr>
<td>FVC</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>TLCO</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>KCO</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Improvement/decline in FEV1 &amp; FVC &gt;10%, TLCO and KCO &gt;15% and rate of decline:</td>
<td></td>
<td></td>
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<tr>
<td>Median annual change in lung function</td>
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</tbody>
</table>

There was a significant difference in median rate of decline of TLCO (p=0.036) and KCO (p=0.014) between Quitters and Smokers, with a trend towards a difference for FEV1 (p=0.141) and FVC (p=0.285).

This case series confirms previous findings supporting the importance of smoking cessation which can have a positive impact on lung function. Continued smoking however can result in continued decline at a rate faster than that reported in healthy normals and patients with COPD.

Put on steroids & 100% O2 but no relief. Bronchoscopy showed air bubbles oozing from irregular opening near superior segment of left lower lobe.

Glue was applied to seal bronchial wall leak & to our amaze subcutaneous em-- at attempts (Smokers). Baseline lung function at diagnosis was compared to current values of FEV1, FVC, TLCO and KCO between Quitters and Smokers:

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Smokers (n=8)</th>
<th>Quitters (n=4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1</td>
<td>Improved</td>
<td>Declined</td>
</tr>
<tr>
<td>FVC</td>
<td>–91ml, –2.44%</td>
<td>–89ml, –5.11%</td>
</tr>
<tr>
<td>TLCO</td>
<td>–139%, –1.56%</td>
<td>–143%, +2.53%</td>
</tr>
<tr>
<td>KCO</td>
<td>–0.0154, –1.00%</td>
<td>–0.098, +6.66%</td>
</tr>
</tbody>
</table>

P3667
A retrospective cohort study of interstitial lung diseases in Denmark
Charlotte Hyldegaard, Elisabeth Bendstrup, Ole Hilberg, Department of Respiratory Diseases, Aarhus University Hospital, Aarhus C, Denmark

Introduction: Interstitial lung diseases are a heterogeneous group of diseases with varying degrees of inflammation and fibrosis. Epidemiological data based on the current diagnostic criteria are sparse.

Objectives: The aim of this study was to characterize the distribution of ILD subtypes in a cohort of Danish patients referred to a tertiary referral hospital.

Methods: We included 431 patients diagnosed with interstitial lung disease (sarcoïdosis excluded) and first visit at our department between April 1, 2003 and April 1, 2009. All diagnoses were re-evaluated according to current diagnostic criteria including the 2011 ATS/ERS IPF guidelines. Details on diagnostics, lung function, comorbidity and treatment were recorded at baseline and throughout the follow-up period.

Results: A total of 186 patients were diagnosed with idiopathic interstitial lung diseases. In this group IPF (n=121) was the most common diagnosis (108 definite and 13 probable IPF) followed by NSIP (n=30), and DIP (n=20). Other large groups comprise 11% of cases and have a median age of 50 years. There was a significant difference in median rate of decline of TLCO (p=0.036) and KCO (p=0.014) between Quitters and Smokers, with a trend towards a difference for FEV1 (p=0.141) and FVC (p=0.285).

This case series confirms previous findings supporting the importance of smoking cessation which can have a positive impact on lung function. Continued smoking however can result in continued decline at a rate faster than that reported in healthy normals and patients with COPD.

Spontaneous pneumomediastinum & subcutaneous emphysema in idiopathic pulmonary fibrosis (IPF) with bronchial wall leak
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Spontaneous pneumomediastinum & subcutaneous emphysema in IPF is due to alveolar rupture but air leak due to bronchial rupture is rare. We present unique case of bronchial leak seen on bronchscopy in a patient of IPF. Non traumatic subcutaneous & mediastinal emphysema due to bronchial rupture in IPF is rare & is being reported for the first time. Case report-55 years male admitted with severe progressive dyspnoea & dry cough of 1 month. Massive subcutaneous emphysema was seen on chest, neck & arms. CT thorax showed pneumomediastinum & IFP in lower lobes.