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

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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 those 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, expiratory 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.

P3652

The efficacy of pirfenidone in scleroderma related interstitial lung disease

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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 lung 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 for the prevention of 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.

P3653

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 impacts on non-specific interstitial pneumonia (NSIP) and interstitial pneumonia (IP) associated with collagen vascular diseases (IP-CVD) are not clear.
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 IPF, NSIP and IP-CVD. Clinical diagnosis are IPF(14), NSIP(14), IP associated with scleroderma(3), and rheumatoid arthritis(1). Based on PaO_2 at rest and SpO_2 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 during pirfenidone administration.

Table 1. Disease severity of patients

	PaO_2 at rest (mmHg)	SpO_2 after 6MWT	n
I	>80		9
II	70–79	When<90%, to III	8
III	60-69	When<90%, to IV	4
IV	<60	unnecessary to perform 6MWT	11

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%, 14%, 0% 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.

P3654

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

%	Strongly Disagree	Disagree	Neutral	Agree	Strongly Agree	No Response
As punctual	2	17	12	40	27	2
As convenient	0	8	12	38	40	2
Felt as reassured	2	10	11	48	29	0
Able to ask questions	0	11	12	44	33	0
Understood the information	0	6	6	52	36	0

Discussion: The majority of ILD patients selected for a telephone consultation were at least 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.

P3655

NSIP: A diagnosis?

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Introduction: Nonspecific 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 (28.8%), 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%), sarcoïdosis (n=2, 2.7%), asbestosis (n=1, 1.4%), 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.

P3656

6-minute walk test (6MWT) in patients with idiopathic pulmonary fibrosis (IPF): Confirmation of the minimal clinically important difference (MCID)

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Introduction: The 6MWT is a practical measure of exercise tolerance in patients with IPF. MCID estimates for 6MWT distance (6MWD) in patients with IPF have ranged between 10–58 m [1-3]; we sought to confirm these estimates in an independent cohort of patients with IPF.
Methods: All patients randomized to placebo in the CAPACITY studies were

included in the analysis. Both distribution- and anchor-based methods were used to estimate the MCID for 6MWD. Distribution-based analyses included standard error of measurement (SEM) and effect size (ES); anchor-based analyses used criterion referencing to estimate the difference in 6MWD between those who did and did not experience significant health events.
Results: A total of 345 patients were included in the analysis. The mean (SD) baseline 6MWD was 404.6 m (90.4). The estimated SEM for 6MWD was 37 m (95% CI, 34–40). The estimated ES was 0.32, based on a mean change of 29.2 m from baseline to Wk 48. 6MWD values were significantly different for those who experienced the composite endpoint of hospitalization or death vs. those who did not; the corresponding MCID was 21.7 m (p=0.047).

Table 1. Estimation of the MCID for 6MWD in patients with IPF

Standard Error of Measurement	Mean	SD	ICC	SEM (95% CI)
6MWT Distance, m (N=338)	404.6	90.4	0.83	**37** (34–40)
Effect Size	**Baseline**	**Follow-up***	**Difference**	**Effect Size**
6MWT Distance, m (mean [SD]) (N=296)	410.5 (90.0)	381.3 (120.9)	**-29.2** (79.6)	0.32
Criterion referencing†		**N**	**6MWD, m‡**	**P-Value§**
Hospitalization or Death No Yes Difference		244 94	410.6 (89.7) 388.9 (90.6) **21.7**	0.047

SEM=standard error of measurement
*Assessed at the week 48 study visit
†Comparison of baseline 6MWT distance between patients who did and did not experience selected health events during the subsequent 48-week period.
‡Data are presented as mean (SD)
§ Independent samples t-test

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Conclusions: Analysis of 6MWD data from a large cohort of IPF patients yielded an MCID estimate 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 transbronchail 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 investigate the effectivity of TBLB and bronchoalveloar lavage (BAL) during FBS in patients with newly diagnosed EAA.
Materials: 66 patients with EAA were enrolled into the study, all of them undergo 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 42/66 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%).
Conclusion: Bronchoscopy with TBLB and BAL even without fluoroscopy control is effective and safe way to establish the diagnosis of allergic alveolitis.

P3658
KL-6 compared to LDH as a prognostic factor in Caucasian patients with idiopathic pulmonary fibrosis
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BACKGROUNDS: Idiopathic pulmonary fibrosis (IPF) is a fatal lung disease with a poor prognosis. Little is known about sensitive prognostic markers for IPF.
Aims: The aim of our study was to investigate the significance of KL-6 compared to LDH as a prognostic marker in Caucasian patients with IPF.
Methods: We prospectively collected 79 Caucasian patients with IPF. Serum samples were obtained at enrollment. Serum levels of KL-6 and lactate dehydrogenase (LDH) were measured by ELISA and the correlation between baseline serum levels of the markers and the survival was evaluated.
Results: Median follow-up period was 372 (1-7088) days. Sixteen (20%) patients died during follow-up. There was no significant difference in the baseline serum KL-6 levels between patients who survived and not survived. Receiver operating characteristic (ROC) curve analysis for predicting non-survival showed larger area under the curve for KL-6 than LDH (0.617 and 0.553, respectively). When the cut-off levels of 1300 U/mL for KL-6 and 280 IU/L for LDH were set, sensitivity, specificity and accuracy for predicting non-survival were 81%, 60% and 66% for KL-6 and 56%, 51% and 52% for LDH. In Kaplan-Meier analysis, patients with baseline serum KL-6 level ≥1300 U/mL showed shorter survival compared with patients with baseline serum KL-6 level <1300 U/mL (p=0.02). LDH showed no statistical impact on the prognosis (p=0.20). In multivariate analysis, baseline serum KL-6 level ≥1300 U/mL was an independent predictive factor for poor prognosis (hazard ratio=4.46; p=0.030).
Conclusions: Baseline serum KL-6 level ≥1300 U/mL is an independent predictive factor for poor prognosis in Caucasian patients with IPF.

P3659
Improved and immediate diagnostics in mediastinal sarcoid lymphadenopathy via endobronchial ultrasound and quadruple testing
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Background: Mediastinal lymphadenopathy carries a wide differential diagnosis, including sarcoidosis, Tuberculosis (TB) and malignancy. Endobronchial Ultrasound-guided Transbronchial Nodal 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 2011. 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 22/25 (88%), 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: The elusive nature of the cause of most diffuse lung diseases has led many to postulate 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; 8 with NSIP/NSIP; 13 with AIP/DAD and 4 with ICLF/CLF. The clinical/radiology and histological diagnosis was according with the 2001 ATS/ERS consensus. Microarray samples was analyzing by immunohistochymistry for measles virus (MV), cytomegalovirus (CMV), hepatitis-C virus (HCV), adenovirus (ADV), respiratory syncytial virus (RSV), Epstein-Barr virus (EBV),

herpes I and II viruses (HVI and HVII). We also correlated sex, age, AIDS and immunosuppression therapy.
Results: We detected by immunohistochemistry the epithelial alveolar infection of MV and CMV in 30.8% and 15.4% in DAD pattern, respectively. Endothelial CMV infection was observed in 25% of CLF pattern. When we divided the age in two groups' patients with ≤ of 43 years old and ≥ of 72 years old, the first group had more infection of MV, CMV than the second group. The Spearman's correlation showed a positive correlation between epithelial and endothelial CMV infection and AIDS (r=0.472; p=0.003).
Conclusion: The epithelial and endothelial viral infections observed in these diseases reinforce the possible viral participation in the pulmonary fibrosis disease. These findings are particularly relevant given the increased interest in the epithelial injury and repair as it relates to the pathogenesis of many diffuse lung diseases.
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P3661
Pirfenidone, proton pump inhibitor, N acetyl cysteine (PINPOINT) therapy for IPF: Tolerance and safety profile among Indian patients
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Rationale: To our knowledge there are no data of PIrfeNidone, PrOton pump Inhibitor, N acetyl cysteine Therapy(PINPOINT) in IPF. 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 liver function tests were performed. Lung function & 6MWT were possible in 25 patients. Patients were followed at 2 weeks, then monthly/quarterly. Prednisolone was administered 10 mg/day & reduced 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 - one 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 scleroderma
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Background: Interstitial lung disease (ILD) is a frequent complication and a leading cause of morbidity and mortality in Systemic Scleroderma (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 SSc.
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 epidemiological, clinical, laboratory features and outcomes between the two groups. The data were compared by the Chi squared correct by fisher exact test and student test.
Results: Thirteen 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 dyspnoea on exertion in 76.9% and dry cough in 46.1%. The ILD was asymptomatic in 3 cases (23%).
The comparison of the two groups showed that digital pitting, inflammatory syndrome 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 study confirms 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 with 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 TLCO 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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Aim: A retrospective study based 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 were studied, based on cinico-pathological and HRCT findings. A number of 12 cases who were undertaken surgury biopsy (OLB/VATS) were selected for the study.
Results: The above 12 patients developed during the follow up period CTD. six cases have f-NSIP on the background and 6 have UIP-like pattern.
Conclusions: The ILD cases without clinical history of CTD and with UIP-like or NSIP pattern on the background of histology speciments, 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 subgroups of patients the prognosis is better than UIP/IPF patients.

P3665
Pulmonary functions in systemic sclerosis
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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 perform 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 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.

P3666

Analysis of cases from an interstitial lung disease (ILD) MDT in a teaching hospital in the West Midlands: Patients with smoking-related disease comprise 11% of cases and have a median age of 50 years

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Introduction: University Hospitals Coventry and Warwickshire is a large acute teaching NHS trust which serves a population of over 1 million. There has been an ILD MDT since 2006. In July 2010 a consultant with an interest in ILD was appointed. The MDT constitutes a thoracic surgeon, pathologist and radiologist with expressed interest in ILD.
Aim: To describe the spectrum of ILD presenting in secondary care.
Method: It is departmental policy to discuss all incident cases of ILD in the MDT. Diagnoses were made according to BTS guidelines, and by consensus. Numbers were compared by $\chi 2$, and ages by Mann-Whitney U-test.
Results: From September 2010 to July 2011, 89 suspected cases were discussed in the ILD MDT, 10 of whom proved not to have significant ILD. The diagnoses of the remaining 79 are given in table below.

Diagnosis	Number	Median Age	Range
Hypersensitivity Pnuemonitis	2	57.5	46-69
Idiopathic Pulmonary fibrosis (IPF)	12	77	57-81
Occupational	4	81.5	71-88
Other Idiopathic Interstial Pnuemonia	33	66	44-84
Rheumatological	19	63	40-83
Smoking Related	9	50	35-70

Definite IPF made up 15% of the cohort, with a median age of 77. Smoking related ILD – Respiratory Bronchiolitis associated ILD and Desquamative Interstitial Pneumonia – account for (*11%*) of cases. The median age of diagnosis is 27 years younger than those with IPF ($p<0.001$).
Conclusion: Smoking related ILD makes a significant proportion of cases discussed at the ILD MDT. Patients are significantly younger than those with IPF. This may reflect an increasing awareness of the radiological and pathological appearance of these diseases.

P3667

A retrospective cohort study of interstitial lung diseases in Denmark

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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 (sarcoidosis 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 were unclassifiable fibrosis (n=62), end stage fibrosis (n=43), extrinsic allergic alveolitis (n=32) and ILD in collagen vascular disease (n=54). Of patients with CVD-ILD 14 showed a UIP pattern, 22 a NSIP pattern and 18 an unclassifiable fibrosis pattern. The remaining 54 patients in the cohort had 15 different diagnoses. Mean age at diagnosis was 61 years (SD=14), and mean symptom duration before referral was 2.5 years. Thoracoscopic biopsy was performed in 39%. The estimated incidence rate is 4.1 per 100,000/year.
Conclusion: The preliminary results of the study of this well-characterised cohort show that IPF is the most common diagnosis, and that a definite IPF-diagnosis can be made in the majority of these cases.

P3668

Respiratory bronchiolitis interstitial lung disease – What is the natural history?

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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 follow up ranging from 8 months to 8 years. All patients were active smokers at diagnosis 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:

Change in lung function

Parameters	Smokers (n=8)			Quitters (n=4)		
	Improved	Declined	Stable	Improved	Declined	Stable
FVC	1	4	3	0	0	4
FEV1	0	4	4	1	0	3
TLCO	0	4	4	1	0	3
KCO	1	2	5	1	0	3

Improvement/decline in FEV1 and FVC >10%, TLCO and KCO >15%.

and rate of decline:

Median annual change in lung function

Parameters	Smokers (n=8)	Quitters (n=4)
FVC	–91ml, –2.44%	–58ml, –1.51%
FEV1	–93ml, –3.03%	+14ml, +2.53%
TLCO	–0.149, –1.56%	+0.216, +2.53%
KCO	–0.0154, –1.00%	+0.098, +6.66%

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.

P3669

Spontaneous pneumomedistinum & 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 bronchoscopy in a patient of IPF. Non traumatic subcutaneous & medistinal 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 pneumomedistinum & IPF in lower lobes.

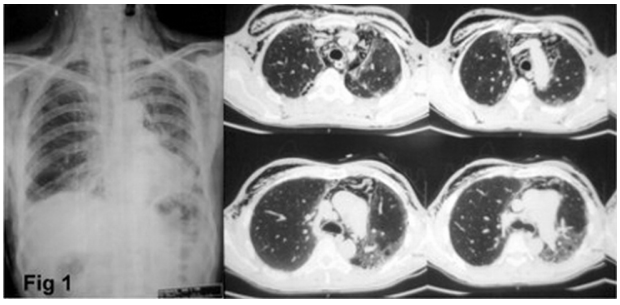
Fig 1

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

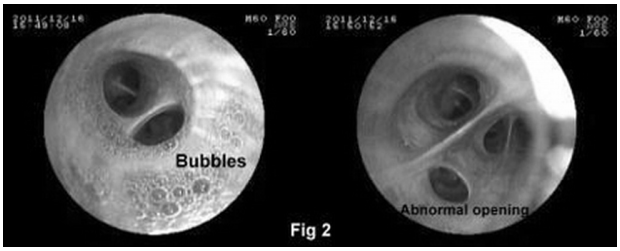

Fig 2

Glue was applied to seal bronchial wall leak & to our amaze subcutaneous emphysema started regressing with clinical improvement. Repeat CT scan showed decrease in mediastinal air. Discussion-Bloomberg considered the cause of non traumatic subcutaneous & mediastinal emphysema to be due to weakness of either alveolar or bronchial wall. Air escapes via bronchovascular channels to mediastinum & subcutaneous tissues, described as Maclin effect.