388. Diffuse parenchymal lung disease II

P3630

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

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Introduction: Exposure to avian proteins is a frequent cause of Hypersensitivity Pneumonitis (HP). We describe a series of NH secondary to exposure to feather duvet.
Material and methods: In the outpatient clinic of interstitial lung disease during the years 2004-2011, 28 patients were diagnosed with HP with definite causality from exposure to feathers contained in the feather duvet. Diagnosis criteria: diagnosis of NH, contact with feather duvet coincident with the onset of symptoms; IgG + and/or positive bronchial challenge test (BCT). In neither case had history of exposure to any agent known as a producer of NH.
Results: 15 male; mean age 59 years. Presentation was acute in 5, subacute in 4 and chronic in 19 cases. Auscultation was normal in 10 patients, revealed crackles in 17 and wheezing in 1. FVC mean was 67.57%, DLCO 52.60%. IgG+ in front of avian antigens in 11/24, and in front of fungi in 16/24. A culture of feather duvet was positive for fungi in 6 cases. The BCT was positive in 8/11 in front of avian antigens and 4/7 against fungi. Pathological study was performed in 16/28 patients, showing a characteristic pattern of subacute NH in 6/16, consistent with NH in 8/16 and UIP pattern in 2/16. Surgical lung biopsy (BPQ) was performed during the study in 7 cases and revised from the samples of another center in 7. During the follow up lung transplantation, was performed in 4 patients (2 of them already had previous BPQ).
Conclusions: Our study confirms that exposure to a minimum but persistent agent may be sufficient for disease development. Diagnosis at an advanced stage of chronic NH is common. Project funded by FIS PI1001577 (ISCIII) and SEPAR 2010.

P3631

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

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Background and objective: Bronchiolitis obliterans organizing pneumonia (BOOP) is a distinct clinicopathological entity histologically characterized by intra-alveolar granulation tissue and absence of extensive fibrotic lesions. Effective macrolide treatment of BOOP has been reported anecdotally. This study aimed to investigate whether alveolar macrophages (AMs) produce aberrant proinflammatory cytokines in BOOP and whether this can be inhibited by clarithromycin (CAM) or azithromycin (AZM).
Methods: AMs collected by bronchoalveolar lavage (BAL) from 6 BOOP patients and 8 non-ILD controls were cultured for 24 hours in the presence or absence of CAM, AZM, lipopolysaccharide (LPS), or dexamethasone (DEX). Tumor necrosis factor alpha (TNF-α), soluble TNF receptor 1 (sTNFR1), sTNFR2, interleukin 1beta (IL-1β), IL-6, IL-8, IL-10, interferon gamma inducible protein 10 (IP-10) and CC chemokine ligand 18 (CCL18) were measured in the culture supernatant by ELISA.
Results: The spontaneous and LPS-stimulated production of all investigated cytokines by AMs was significantly increased in BOOP compared to controls. CAM and AZM induced a dose-dependent suppression of spontaneous TNF-α, sTNFR2, IL-6, IL-8 and CCL18 production ($p < 0.05$). CAM also inhibited the IL-1βproduction. CAM and AZM significantly and dose-dependently attenuated the LPS-stimulated production of sTNFR1, sTNFR2, IL-8 and CCL18 ($p < 0.05$). CAM also inhibited the LPS-stimulated TNF-α, IL-1β, IL-6 and IL-10 production.
Conclusions: AMs from BOOP patients produce abundant proinflammatory cytokines which may be pivotal in the disease pathogenesis. Macrolides inhibit this cytokine production, CAM more efficiently than AZM.

P3632

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

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Purpose: Patients with combined pulmonary fibrosis and emphysema syndrome (CPFE) and those with interstitial pneumonia (IP) have a high risk of developing lung cancer. The present study compared the clinical features of patients with lung cancer and CPFE with those of patients with lung cancer and IP.
Methods: 48 patients with lung cancer and CPFE (CPFE group) and 30 patients with lung cancer and IP (IP group) had surgery and were included this study.
Results: All patients in the CPFE group and 24 of 30 patients in the IP group were current or ex-smokers (p=0.01) (Table 1). The CPFE group tended to have more patients with IPF than the IP group (p=0.09). FEV1.0% predicted was significantly lower in the CPFE group than in the IP group (Table 2). Acute exacerbations of

Table 1. Patients characteristics

		Patients with lung cancer and CPFE (n = 48)	Patients with lung cancer and IP (n = 30)	p value
Sex	Male	45	20	0.02
	Female	3	10	
Age	Median (Range)	70 (30-82)	73 (58-83)	0.05
Smoking	Smokers	48	24	0.01
	Nonsmokers	0	6	
Histology of lung cancer	Adenocarcinoma	18	19	0.19
	Squamous cell carcinoma	18	13	0.81
	Large cell neuroendocrine carcinoma	5	1	0.17
	Adenosquamous	3	1	0.44
Types of IP	Idiopathic interstitial pneumonia			
	IPF	36	17	0.09
	Fibrosing NSIP	1	1	0.73
	DIP	0	2	0.07
	Unclassified	9	7	0.63
	Collagen vascular disease	2	3	0.31

interstitial lung disease after surgery developed in three patients (6.3%), one of whom died, in the CPFE group and two patients (6.7%) in the IP group (p=0.94).

Table 2. Laboratory findings and pulmonary function tests

		Patients with lung cancer and CPFE (n = 48)	Patients with lung cancer and IP (n = 30)	p value
Blood gas analysis before operation	PaO_2 (Torr)	85.0±11.3	85.0±10.8	0.96
	A-aDO_2	13.1±10.9	15.2±11.5	0.84
Laboratory Findings	KL-6 (U/ml)	536.0±549.0	766.0±658.2	0.39
	SP-D (μg/l)	114.4±79.3	128.5±74.7	1.00
	SP-A (ng/l)	56.3±30.4	59.7±18.7	0.72
	LDH (U/l)	162.0±34.5	168±38.6	0.66
	CRP (mg/dl)	0.2±1.5	0.2±0.4	0.07
Pulmonary function tests	VC % pred	102.0±15.7	99.5±16.3	0.28
	FEV1.0 % pred	93.5±16.4	99.0±11.2	**0.04**
	TLC % pred	98.5±14.2	98.0±8.6	0.60
	RV % pred	84.5±26.2	93.0±21.1	0.82
	DLCO % pred	63.0±13.8	80.5±11.0	0.16

Data are presented as mean ±SD

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

P3633

A case of lymphoid interstitial pneumonia associated with common variable immunodeficiency

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Introduction: Lymphoid interstitial pneumonia (LIP) represents a rare disease that typically occurs in association with autoimmune diseases and dysproteinemia. We report a case of LIP in a patient with common variable immunodeficiency (CVID).
Case report: In October 2009, a 51 year old woman presented with a 12-months history of recurrent pyrexia and was subsequently diagnosed with CVID. CT scans showed bilateral pulmonary consolidations, while bronchoscopy revealed acute inflammation of the bronchial mucosa and purulent secretion. The patient received antibiotic treatment and immunoglobulin replacement. She remained asymptomatic until January 2012, when she was readmitted with increasing dyspnea and fatigue for 2 weeks prior to admission. CT scans showed progressive opacities mainly in the lower zones. Despite immediate treatment with broad-spectrum antibiotics, the patient developed acute respiratory failure requiring invasive mandatory ventilation and, ultimately, extracorporeal membrane oxygenation. Open lung biopsy was performed 5 days after admission. Histologic workup showed diffuse interstitial infiltration of T lymphocytes, plasma cells and histiocytes, consistent with a diagnosis of LIP. Treatment with prednisone (1mg/kg) resulted in rapid clinical and radiological improvement. The patient was discharged 4 weeks after admission. At present, she remains clinically stable at a steroid dose of 0.25mg/kg.
Discussion: The finding of bilateral ground-glass infiltrates in a patient with CVID should raise suspicion of LIP. Open lung biopsy is required to establish the diagnosis. Treatment is primarily based on corticosteroids, which resulted in excellent response in this patient.

P3634

Disease progression according to IPF phenotype

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Introduction: Our understanding of disease progression in IPF is based on cohorts of patients with 'definite' IPF. Such studies have reported a prognostic index for IPF (Du Bois 2011 AJRCCM 184; 459). In practice a diagnosis of definite IPF is not always attained; in some cases where the HRCT pattern is not incontrovertibly UIP, corroboratory invasive investigations are not performed. We define this condition 'probable IPF'.
Aims: To determine 1) disease progression and 2) the prognostic value of an IPF index in definite or probable IPF.
Methods: Consecutively presenting patients with IPF were prospectively recruited to a database. All IPF diagnoses required an HRCT appearance of UIP with ≥ 70% probability. Patients with HRCT scans with ≥ 95% probability of UIP, or a UIP biopsy were defined as definite IPF. BAL was not included in the diagnostic criteria.
Results: Of 193 patients, 89 had definite IPF. Median survival for definite v probable IPF was 3.2 v 6.0 years respectively (HR 1.45; 95%CI 0.94 to 2.25, p=0.09 adjusting for age, sex, height, VC and smoking). A relative decline in VC of 5-10% in the first 6 months was associated with increased risk of death (HR 3.12; 95% CI 1.55 to 6.30). Similar results were obtained in patients with definite and probable IPF. An IPF index was calculated in our cohort. Scores of 0-8 (n=97), 10-29 (n=76) and 30-61 (n=20) predicted median survival of 74, 48 (HR 2.02; 95%CI 1.26 to 3.23) and 12 months (HR 15.3; 95%CI 8.22 to 28.4) respectively ($p < 0.001$). The index had similar predictive value in definite and probable IPF.
Conclusions: Patients with both definite and probable IPF have poor prognoses but survival was worse in definite IPF. The IPF index may be of clinical value in definite and probable IPF.

P3635

Sarcoidosis and tuberculosis: A rare combination?

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Tuberculosis and sarcoidosis are chronic granulomatous diseases that are similar in many aspects, although different. They occur concomitantly very rarely. TB is an infectious disease caused by M. *tuberculosis* morphologically defined by granulomas with caseous necrosis. Sarcoidosis is a systemic disease of unknown etiology, and is characterized by noncaseous granulomas.
Aim of the study: To evaluate weather the high incidence of TB in our country affects the diagnosis and management of sarcoidosis.
Material and methods: We performed an analysis of 97 patient's clinical records with biopsy proven or highly suggestive of sarcoidosis, cases registered in a third level medical institution, during year 2011.
Results: We found 3 (3,1%) patients with concomitant diagnosis of TB and sarcoidosis, in whom sputum culture revealed M. *tuberculosis* (1-MDR tuberculosis, 1- poly-resistant). After 2 months of standard anti-TB treatment we have obtained culture conversion, but no improvement in chest X-ray and HRCT. Supplementation with corticosteroids (CS) leaded to full resorption of pulmonary lesions. Other 8 (8,2%) patients from 97, microbiologically negative, where initially diagnosed with pulmonary TB based on histological examination (caseating granulomas). Anti-TB treatment inefficiency imposed reconsideration of the diagnosis and initiation of steroid treatment, with clinical and radiological improvement after 4 months of CS.
Conclusion: Infection with *M. tuberculosis*, seems to be not so infrequent in patients with sarcoidosis. The finding of necrotizing granulomas alone is not sufficient for the final clinical diagnosis of tuberculosis. Due to high incidence of tuberculosis in our country there is a tendency to overdiagnose tuberculosis.

P3636

Precocious detection of pulmonary hypertension in idiopathic pulmonary fibrosis

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The aim of our study is:(1)to evaluate the prevalence of Pulmonary Hypertension (PH) in Idiopathic pulmonary fibrosis (IPF) patients (pts);(2)to determine the accuracy of PAPs measurement with Doppler echocardiography (DE) versus Right Heart Catheterization (RHC);(3)to analyze survival in pts with or without PH;(4)to determine any correlation between functional data and haemodynamic parameters. We analyzed 54 IPF pts referred to our Centre who underwent RHC. PH was defined when PAPm ≥25 mmHg. PH was diagnosed in 30/54 pts (55%, PAPm=30±5mmHg, PAPs=48±10, PW=11±3, RVP=3,1±2). There was a significant correlation between PAPs evaluated at DE and RHC ($p<0,0001$), but Bland-Altman analysis illustrated mean difference: 5,1±22,4; the large limits of agreement (-17 +27) showed that DE should not be used to replace RHC. No difference in survival was found between PH group(1) and no PH group(2). Pts were divided according PH onset [<12 months (m) from IPF diagnosis: group A, or >12 m: group B] and we observed a significant difference in survival between group A and B (p=0,0042, 35m versus 56), between group A and all other pts(group 2+B; p=0,0095, 35 versus 52).11(91%) of group A needed oxygen-therapy after a median time of 8m since IPF diagnosis, while 32(76%) of all other pts (group 2+B) after 16m(p=0.029). We observed a significant correlation between Six Min Walking test distance (WTD) performed in room air and PAPs at RHC(p=0.039). Our data confirms that PH has an high prevalence in IPF and RHC is the gold standard for PH diagnosis. An early onset of PH is related to a worst prognosis and to a precocious oxygen-therapy request. A decrease of WTD could help clinicians to suspect PH in IPF pts.

P3637

Obstructive findings in sarcoidosis, diagnosis and therapies modalities

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Introduction: Symptoms of respiratory disturbances are common during the

course of sarcoidosis. The aim of analysis is to notifie obstructive findings in sarcoidosis.
Method: The analyses is restrospective in all incominig patients to the Clinic for lung diseases and tuberculosis CC of Serbia in Belgrade.
Results: 127 (8,7%) patients with dry cough and occasionaly dyspnea had the impairent of lung function. The main findings were: FEVl/VC 63,1% in 57-44,8% patients; in 27-21,1% patients – FEVl/FVC were 55,7%, in 3-2,3% patients FEVl/FVC were 68.7%. Gender distribution were as followed: 264 (66,2%)Female-Male 135 (33,8%); average ages were 43,32 years. Extrathoracis sarcoidosis involvement were find out in 3 (2.3%) patients (skin, lymph nodes,).Lung and nonpulmonary sarcoidosis is notify in 27 pts and in 97 pts lung were the only sarcoidosis manifestation. Radiographic stadium of lung involvement: I-87(68.5%), II-31(24,4%), III-7%(5,%), IV-1 (0,78%). Acute onset of sarcoidosis is predominatly with average level of ACE: 75.3 U/L. After obtaining the diagnosis of sarcoidosis, inhaled corticosteroides were administered, 160 mcg-daily through 3 months with controls which meansa: ACE and UCa/24h levels, lung function, chest X ray. Regression of symptoms such as dry cough were obtained in 55% patients after the period of 2 months; 45% patients were excluded due to persistence of symptoms seeking prednisone.
Conclusion: Inhaled corticosteroid have the role in sarcoidosis treatment resolving symptoms such as cough, improvement of FEV1, but the main therapy in most of the patients is still orally administired prednisone.

P3638
Natural history of idiopathic pulmonary fibrosis: Are slowly progressive and rapidly progressive really steady conditions?
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Idiopathic pulmonary fibrosis (IPF) is a devastating lung disease with heterogeneous clinical course. Some patients experience an accelerated disease progression (rapid progressors) while other remain relatively stable over time (slow progressors).
The aim was to investigate the different course of the disease in relation to survival. The study population included 55 IPF patients (age at diagnosis 53±1) categorized in rapid progressors and slow progressors by two distinct criteria: pre-diagnosis criteria (time from symptoms onset and IPF diagnosis) or by post-diagnosis criteria (decline in FVC%pr. over 12 months). When stratified by pre-diagnosis criteria 18% were rapid progressors while 66% were slow ones.When stratified according to post-diagnosis criteria 67% were rapid progressors and 33% were slow ones. The coefficient of agreement between the two criteria was 70% and 75% for slow and rapid progressors respectively indicating that up to 30% of patients did not maintain the same label. Stratification by pre-diagnosis criteria was not related to survival. Conversely, stratification by post-diagnosis criteria had a prognostic significance; indeed, rapid progressors had decreased survival as compared to slow ones (28±1Vs49±8mo.,p=0.02). Of interest, rapid progressors according to post-diagnosis criteria, often display an unstable decline alternating periods of functional stability to a rapid deterioration.
In conclusion our data suggest the need to be cautious in labelling IPF patients to a fixed phenotype from the beginning of symptoms till death. It is possible that IPF patients show a variable and unpredictable clinical course rather than a steady condition.

P3639
The clinical relevance of autoimmunity in idiopathic pulmonary fibrosis
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The aim of this study is to evaluate the clinical significance of autoimmunity in IPF.
This is a retrospective controlled study comparing clinical characteristics and outcome of IPF patients with (n=73) and without (n=87) positive autoimmunity.
Seventy-three cases with positive autoimmunity included: 46 (63%) ANA (anti nuclear antibody), 20 (27%) RF (rheumatoid factor), 2 (3%) anti CCP (anti cyclic-citrullinated), 3 (4%) ENA (antibodies to extractable nuclear antigens), 2 (3%) ANCA (anti-neutrophil cytoplasmic antibody).
No differences were found between patients with positive and negative autoimmunity in terms of age, gender, smoking history, prevalence of familial IPF, high resolution computed tomography features.

Characteristics of IPF patients

	IPF-positive autoimmunity	IPF-negative autoimmunity
Age, median (range)	60 (47–9)	64 (39–84)
Male gender, n (%)	48 (66)	66 (76)
Smoking, current and former smokers, n (%)	58 (79)	59 (68)
Familial – IPF, n (%)	16 (22)	16 (18)
HRCT features consistent with UIP, n (%)	26 (36)	32 (37)
Median follow up, months (range)	32.19 (0.68–106.9)	32.48 (0.42–114.3)
AZA, CSS, NAC treatment, n (%)	9 (12)	15 (17)

During follow up two patients developed an autoimmune disease (one rheumatoid arthritis and one scleroderma).
Survival between IPF patients with and without positive autoimmunity did not differ 77.4 months (95% CI 56.1-97.8).
Conclusion: A small minority (2.7%) of IPF patients with positive autoimmunity test at diagnosis developed an autoimmune disease during follow-up. The presence of positive autoimmunity testing do not influence neither IPF clinical presentation nor survival.

P3640
Sarcoidosis and autoimmune thyroid disease
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Background: Association between Sarcoidosis and other autoimmune diseases has been previously described and a common pathogenesis has been hypothesized.
Objectives: A descriptive retrospective study has been conducted to evaluate the incidence of thyroid diseases in patients with pulmonary sarcoidosis.
Methods: We conducted a retrospective chart review of all patients diagnosed with sarcoidosis between 2004 and 2011 at the Morgagni Hospital, Forlì; those who were also diagnosed as having an autoimmune thyroid disease were selected.
Results: 39 out of 246 (15,9%) patients with sarcoidosis were identified as having autoimmune thyroid disease, 9 male and 30 female. Sarcoidosis presented as Löfgren's syndrome in 6 patients patients (15,4%). 3 patients developed Graves' disease (7.7%), 12 patients developed Hashimoto's thyroiditis with hypothyroidism (30.8%) and 24 patients had thyroid nodules with normal thyroid function (61.5%).
Conclusions: Our study suggests that sarcoidosis may be associated with autoimmune thyroid diseases at some time of its evolution, either as hyperthyroidism or hypothyroidism. We are now following up sarcoidosis patients with no thyroid diseases to see if any thyroid disease can develop after the diagnosis or during the period of activity of sarcoidosis.

P3641
Assessment of disease severity using imaging scores in pulmonary sarcoidosis
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The value of imagistic methods in diagnosing and assessing the inflammatory activity in sarcoidosis is well known.
The aim was to assess the association between imagistic findings (radiographic score, and HRCT score by Oberstein *et. al.*[SarcoidosisVasc Diffuse Lung Dis 1997,14:65–72]) and disease severity defined by functional disturbances in sarcoidosis.
We have evaluated the clinical records of 70 patients with biopsy proven or highly suggestive of sarcoidosis, cases registered in a 3rd level medical institution, during year 2011, who underwent chest X-ray, HRCT, and pulmonary function tests. The HRCT images were scored by two readers. Spearman's rank correlation coefficients were calculated to estimate the association between imagistic scores and respiratory function disturbances.
We found a relationship between both radiological staging and HRCT abnormalities with functional parameters.
All HRCT subscores, except lymph node enlargement, correlated with FEV1, FVC, $MMEF_{25-75}$, and DLCO ($p<0,001$).
Compared with radiological stages, HRCT findings appeared to be more sensitive in tracing abnormal gas exchange, with no significant difference in other functional parameters.

Abstract P3641 – Table 1. Correlations between radiographic stages, HRCT scores and subscores and functional parameters

	Thickening of the bronchovascular bundle	Parenchymal consolidation	Intraparenchymal nodules	Septal/non-septal lines	Focal pleural thickening	Total HRCTscore	Radiographic score
FEV1	–0,34**	–0,16	–0,12	–0,38**	–0,31**	–0,37**	–0,38**
FVC	–0,23	–0,19	–0,13	–0,29*	–0,32**	–0,31*	–0,31**
DLCO	–0,26*	–0,41**	–0,36**	–0,53**	–0,12	–0,52 **	–0,38**
MMEF25–75	–0,21	–0,17	–0,27*	–0,31**	–0,26*	–0,31**	–0,34**
TLC	–0,13	–0,07	0,02	–0,16	–0,26*	–0,23*	–0,12

*p<0,05; **p<0,001.

P3642

Myocardial mechanics for the early detection of cardiac sarcoidosis

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Background: Speckle tracking has emerged as valuable tool for comprehensive assessment of regional myocardial function, providing angle-independent measurements of ventricular strain. Aim of this study was to evaluate left ventricular (LV) function in patients with newly diagnosed sarcoidosis, utilizing the novel method of 2D speckle tracking.
Methods: Forty one patients with sarcoidosis and unremarkable medical history of cardiovascular disease, as well as 20 healthy age- and gender-matched controls underwent echocardiographic study. Apical 4-,2-,3- chamber as well as short axis acquisitions were made. In addition to conventional 2D, Doppler and TDI measurements, speckle tracking echocardiography was applied and LV global longitudinal strain was derived from the obtained images. Moreover, LV base and apex rotation angles were assessed from which LV twist was derived.
Results: The mean age of patients (17 men) was 41±6.5 years old. Compared with controls, patients had similar conventional 2D and Doppler measurements. TDI revealed increased E/E' in the patient group vs control group (8.72±1.65 vs 4.6±1.32, $p<0.05$). Strain analysis demonstrated reduced global longitudinal strain values in the patient vs control group (18.86±1.79% vs 21.88±2.18%, $p<0.05$). Furthermore, LV twist was increased in the patient group as compared to the healthy individuals (12.5±2.6° vs 10.2±1.8o, $p<0.05$).
Conclusions: Speckle tracking echocardiography revealed impaired strain and rotational indices, implying elevated filling pressures of the left ventricle. This may represent a sign of myocardial involvement in patients with sarcoidosis. Therefore deformation imaging could be valuable adjunct for screening.

P3643

Overexpression of angiotensin II type 1 receptor (AGTR1) and lymphatic vasculature rarefaction is present in scleroderma with pulmonary compromised

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Background: The aim of this study was to evaluate the activity of angiotensin II type 1 and 2 receptors and lymphatic vessels in lungs of patients with systemic sclerosis comparing to normal lung tissue (NLT).
Methods: Lung specimens were obtained from 23 females patients with scleroderma. NLT were obtained from 10 individuals who's died suddenly of non-pulmonary causes. Immunohistochemistry and histomorphometry were used to evaluate the amount of AGTR-1 and AGTR-2 pulmonary expression. We also studied the lymphatic (D2-40) expression and the percent of area occupied by these lymphatics in lung specimens. Pulmonary fibrosis was obtained from high-resolution computed tomography (HRCT) score.
Results: We observed higher amount of AGTR-1 and AGTR-2 expression in the pulmonary parenchyma of patients with scleroderma when compared with NLT ($p>0.01$). The density of lymphatic vessels was markedly reduced in pulmonary scleroderma when compared with NLT ($p=0.02$) group. Similar situation was observed when we compared the area occupied by these lymphatics ($p=0.001$) group. Patients with several fibrosis showed a increased expression of AGTR-1 and increased area occupied by lymphatic than minimal fibrosis ($p<0.05$) patients.
Conclusions: Our data showed that the AGTR-1 and AGTR-2 and lymphatic system are very affected in pulmonary SSc. AGTR-1 showed higher expression in more pulmonary fibrosis HRCT score. Rarefaction of lymphatic vasculature was correlated with degree of pulmonary fibrosis and pulmonary function test. The role of AGTR-1 and lymphatics are probably essential in evaluation and progression of Systemic Sclerosis.
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P3644

The relationship between the shuttle walk test, lung function and BMI in patients with sarcoidosis

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Introduction: Monitoring patients with sarcoidosis is important and challenging. The incremental shuttle walk test is used in outpatient settings and we assessed its correlation with lung function, BMI and age.
Method: A retrospective study of data from 63 patients with sarcoidosis who had had a shuttle walk with their lung function tests. Spearman's correlation coefficient r_s was used to assess the relationship between the shuttle walk (distance walked, oxygen saturations) and Borg score (before and after exercise), with lung function (FEV1, FVC and DLCO[% predicted]), age and BMI. A stepwise multiple linear regression model of distance walked was developed.
Results: Correlation is seen between the shuttle walk and lung function.(table1)The distance walked also correlated with age (r_s=-.372, p=.003) and BMI (r_s=-.303, p=.016). There was correlation between the Borg score post exercise and the BMI (r_s=.253, p=.046). The multiple regression model of distance walked identified BMI (t=-3.195, p=.002), age (t=-2.963, p=.002) and FEV1 (t=3.061, p=.003) as significant independent variables.

Correlation between shuttle walk test and lung function

	FEV1%		FVC%		DLCO%	
	r_s	p Value	r_s	p Value	r_s	p value
Distance	0.367	0.003	0.325	0.009	0.332	0.009
Resting saturations	0.364	0.003	0.411	0.001	0.392	0.002
Saturations post exercise	0.454	0.000	0.550	0.000	0.641	0.000
Change in saturation	–0.309	0.014	–0.425	0.001	–0.561	0.000
Resting Borg score	–0.397	0.001	–0.330	0.008	0.339	0.007
Borg score post exercise	–0.285	0.024	–0.231	0.068	0.276	0.031

r_s Spearman's correlation coefficient.

Conclusion: The shuttle walk test may be useful in monitoring patients with sarcoidosis as it correlates with lung function. The influence of BMI on distance walked cannot be underestimated.

P3645

Interstitial lung disease associated with autoimmune thyroiditis (ILD-AT)

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Aim. To show some peculiarities of an orphan syndrome which is often misdiagnosed on practice.
Patients and methods. 8 cases of ILD-AT were revealed among about 5000 patients with various interstitial lung diseases during 25 years.
Results. ILD-AT was diagnosed in 7 female patients (age 49-60 yrs) and 1 male (age 23 yrs). In 5 cases the onset of pulmonary and thyroid disease was simultaneous in 3 autoimmune thyroiditis (AT) appeared several years earlier than ILD. The average duration of disease was 8.6 yrs to the moment of ILD-AT was diagnosed (range 1-27 yrs). In all patients ILD-AT presented with constant dry cough, dyspnea and crackles in lungs. NSIP pattern was seen on HRCT scans of all patients. Spirometry found out restriction in 6 of 8 cases (FVC 52-20%) while FEV1/FVC ratio was not decreased and bronchodilator test was negative. DLCO was lowered up to 52.6-25.2% as well as PaO_2 arterial blood level (69-49 mm Hg) in all patients. Long term treatment with low dose corticosteroids (during 2-27 yrs) prevented disease progression. The level of thyroid peroxydase in serum initially elevated in all of patients returned to normal after treatment. The results of treatment were as much better as shorter was the disease duration.
Conclusions. Appearance of constant dry cough and dyspnea should be the reason for chest HRCT, spirometry and DLCO measurement in patients with AT. Patients with interstitial lung disease of unknown origin should be tested for AT. In spite of rather favorable course ILD-AT should be diagnosed in time and treated long to avoid diffuse pulmonary fibrosis formation.

P3646

6-minute walk test predicts pulmonary artery pressure in patients with collagen vascular disease associated interstitial pneumonia

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Objective: It has been reported clinical importance of pulmonary hypertension(PH) in collagen vascular disease(CVD), however the role of PH in interstitial pneumonia related CVD(CVD-IP) has been scarcely evaluated. We sought to determine the prediction factor of mean pulmonary artery pressure(MPAP) in patients with CVD-IP.
Method: Patients with CVD-IP underwent right heart catheterization(RHC) within 3 months of initial evaluation at our institution. Patients with left ventricular dysfunction, incomplete follow-up, and patients with respiratory failure were excluded.
Results: We studied 44 patients, (19 male, mean age 59.8±11.0years). They were 13 with RA, 13 with SSc, 9 with PM/DM, and 9 with others. PaO2 at rest was 83.1±9.6mmHg,MPAP was 17.2±5.5mmHg (>20mmHg, 13(29.5%)), cardiac index(CI) was 3.6±0.8L/min/m^2, pulmonary vascular resistance index(PARI) was 223±102 Wood units/m^2, %vital capacity(%VC) was 78.4±21.0%, percentage of carbon monoxide diffusing capacity (%DLco) was 50.9±17.6%, and 6-min walk distance(6MWD) was 502±142 m, and minimum SpO_2 at 6-min walking test ($mSpO_2$) was 85.6±5.9%, respectively. The median observation period was 31.8 months with 11 patients died. In the univariable model, MPAP was significantly correlated with %DLco (r=-0.377,P=0.013), and $mSpO_2$ (r=-0.552, P=0.0001). In the multivariable model, MPAP was significantly correlated with 6MWD (r=-0.360,P=0.019) and $mSpO_2$ (r=-0.595,P<0.001).
Conclusion: 6MWD and $mSpO_2$ were independent predictors of MPAP in CVD-IP.

P3647

Sarcoidosis – Diagnostics, prognosis and therapy in everyday pneumological practice. A retrospective analysis

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Background: Currently, prognostic factors, or therapy in sarcoidosis are frequently discussed, however, up-to-date data concerning the incidence and the management of sarcoidosis in everyday pneumological practice are still missing. Therefore, we decided to analyze the characteristic of comprehensive cohort of sarcoidosis patients.
Methods: We retrospectively analyzed all 169 patients, whom the diagnosis of sarcoidosis was set at our pulmonary department in years 2005-2010.
Results: Median age of sarcoidosis patients was 48 years (20-79). Females:males ratio was 1.5:1. Non smoker:smoker ratio was 2.2:1. Familial occurrence was observed in 4 patients. At diagnosis, stage 0 was present in 3.5% patients, stage I in 34% patients, stage II in 49.5% patients, stage III in 11% patients, and stage IV in 2% patients. In 76 patients there was extrapulmonary sarcoidosis. The coincidence of sarcoidosis with other autoimune diseases was observed in 10 patients; 6 patients developed trombembolic disease. Spontaneous resolution was seen in 36% patients; 57% of stage I, 29% of stage II, and 11% of stage III. 64% received corticosteroids. 16% of patients developed chronic sarcoidosis. In sarcoidosis patients with spontaneous resolution, no relapse of disease was observed. On the other hand, 10% of treated patients relapsed. Median time to sarcoidosis relapse was 6 months. There was no difference in age between good and poor risk groups of patients. Observed lethality was 1.2% (2 patients).
Conclusion: Male gender, X-ray stage I or II, lymphocytic alveolitis in bronchoalveolar fluid, spontaneous regression, and active disease duration up to 2 years appear to be good prognostic factors.

P3648

BALF cells expression phenotypes and cytokine levels in BALF supernatant in fibrosing interstitial lung diseases

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Aims: Fibrosing interstitial lung diseases (fILDs),mainly idiopathic pulmonary fibrosis (IPF) and fibrotic nonspecific interstitial pneumonia (NSIP), are processes with unknown etiology and grim prognosis. We hypothesize that expression profile of bronchoalveolar lavage fluid (BALF) cells and BALF cytokine concentrations in fILDs depend on Th2 cytokine milieu and enhance fibroproliferative healing.
Methods: 10 patients with fILDs and 9 controls (C) were included into the study. BALFsamples were investigated using FACS to evaluate spontaneous and stimulated production of TNF alpha, percentage of lymphocytes expressing IL-2, IFN-gamma and TNF- alpha and percentage of epithelial cells expressing proteinase-activated receptors (PAR 1,2,3). IFN- gamma, IL-1 beta, IL-2, IL-4, IL-5, IL-6, IL-8, IL-10, IL-12p70 and TNF beta values were evaluated by means of bead based analysis. Concentrations of CD124, PAR-2, TGF-beta 1 and TNF-alpha in BALF were detected by ELISA as well.
Statistical methodology: The differences in basic and derived characteristics were tested via standard two-sample two-sided t-test with correction by False Dicsovery Rate (FDR) methodology.
Results: We observed significantly higher concentrations of IL4R and TGF- beta 1 in BALF in fILDs versus C. The expression of PAR-2 on EP was higher in fILDs compared to C as well.
Conclusions: Our results supported a hypothesis about the role of Th2 type immune response in pathogenesis of fibrotic ILDs. The higher concentrations of IL-4R might be a sign IL-4 influence, and the increased expression of TGF-beta and PAR-2 gives evidence for ongoing epithelial- mesenchymal transition and fibrogenesis in fILDs.

P3649

Pulmonary eosinophilic syndromes characterization. The role of peripheral eosinophilia as a marker of clinical evolution

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Introduction: Pulmonary eosinophilic syndromes (ES) are a heterogeneous, rare and not fully characterized group of diseases.
Objectives: Evaluation of ES patient's clinical features and its evolution.
Methods: 47 patients with >20% BAL eosinophils or compatible histological features were included and its clinical files retrospectively analyzed.
Results: After excluding 19 patients that had other diseases and 9 because of lacking information, 19 patients were enrolled: 8 presented Chronic Eosinophilic Pneumonia (CEP), 6 Churg-Strauss Syndrome (CSS), 3 Acute Eosinophilic Pneumonia and 2 Allergic Bronchopulmonary Aspergillosis (ABPA). Mean age at symptoms onset 37±14.6 and at diagnosis 43±14.3 years. The majority was female (68.4%) and nonsmokers (76.5%). Peripheral eosinophilia was observed in 78.9% patients (1.73±1.32x109/L), mean eosinophilia in BAL was 51.1±29.16%, airflow obstruction was observed in 55.6% patients, 22.2% had restrictive pattern, 16.7% mixed pattern and 5.6% normal values, without significant differences among entities. Concerning CEP, CSS and ABPA cases after treatment, 83.3% presented obstructive pattern, 16.7% normal values and a significant increase of TLC (p=0.038) and RV (p=0.018) were observed. The RV raise was associated with mild peripheral eosinophilia or persistent eosinophilia (p=0.045). Mild peripheral eosinophilia at diagnosis was associated to its persistency after treatment (p=0.023).
Conclusion: In this series, mild peripheral eosinophilia was associated with its persistency and respiratory hyperinflation after treatment, suggesting that the degree of peripheral eosinophilia is associated to a distinct clinical evolution.